Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 3: Middle Ear and Mastoid

Chapter 35: Tumors of the Middle Ear and Mastoid

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Tumors may involve the middle ear, mastoid, and temporal bone primarily, metastatically, or by extension from a contiguous area. The primary neoplasms of the middle ear, mastoid, and temporal bone, not necessarily in the order of their frequency, are glomus jugulare tumors, adenomas, adenocarcinoma, squamous cell carcinoma, fibrosarcoma, neurofibroma of the facial nerve, meningioma, hemangioma, giant cell tumor, osteoma, and acoustic neuroma (Table 1).

Tumors from distant primary foci may metastasize to the middle ear, mastoid, and temporal bone. These include adenocarcinoma of the prostate, mammary carcinoma, hypernephroma or renal carcinoma and other rare lesions, adenoma, bronchogenic carcinoma, gastrointestinal carcinoma, and melanoma. Neoplastic systemic diseases such as leukemia, multiple myeloma, and lymphomas may also involve these structures.

The middle ear, mastoid, and temporal bone occasionally may be invaded by tumors from adjacent areas, such as meningioma, glioma, neurilemmoma, cylindroma of the parotid gland, epidermoid carcinoma and melanoma of the skin of the external auditory canal and auricle, and, finally, nasopharyngeal malignancies.

Occasionally one notes non-neoplastic lesions of bone, xanthomatosis, and fibrous dysplasia, which are mistaken at times for tumors.

Primary Tumors of the Middle Ear, Mastoid, and Temporal Bone

Glomus Jugulare Tumors

Glomus tumors are the most common benign neoplasms of the middle ear. Despite their relative rarity, a great deal of attention has been focused on these tumors because advances in radiographic technique, surgical exposure, vascular control, and combined surgical and radiation therapy have led to improved diagnosis and treatment. Glomus tumors are of particular interest to otolaryngologists because they present a number of unique characteristics. First, although benign histologically, these tumors can grow aggressively and cause multiple cranial nerve abnormalities. Second, the lesion may first develop at a younger age but only manifest itself many years later after slow growth. Third, glomus tumors pose a diagnostic challenge because of their potential to exhibit bilaterality, to occur in multiple family members, to cause endocrine changes, and to be associated with other carotid body tumors. Finally, the location of these tumors at the base of the skull makes them difficult to expose and remove completely.

Table 1. Temporal Bone Neoplasms

I. Primary Tumors
A. Benign
- 1. Glomus jugulare
- 2. Neuroma
  - a. von Recklinghausen's disease
- 3. Meningioma
- 4. Bony lesions
  - a. Osteoma
  - b. Exostoses
- 5. Fibro-osseous tumors
  - a. Ossifying fibroma
  - b. Fibrous dysplasia
  - c. Giant cell tumor
- 6. Choristomas / teratoma
  - a. Salivary gland
  - b. Lipoma

B. Malignant
- 1. Squamous carcinoma
  - a. Verrucous carcinoma
- 2. Adenocarcinoma
  - a. Adenomatous lesions
  - b. Adenocystic carcinoma
- 3. Sarcoma
  - a. Rhabdomyosarcoma
  - b. Fibrosarcoma
  - c. Chondrosarcoma
  - d. Osteogenic sarcoma
  - e. Liposarcoma
- 4. Radiation-induced carcinoma

II. Metastatic Tumors
A. Prostate
B. Breast
C. Gastrointestinal tract
D. Renal cell
E. Lung
F. Multiple myeloma
G. Lymphoma
H. Leukemioa

III. Contiguous Tumor Invasion
A. Neuronal Structures
- 1. Meningioma
- 2. Glioma
- 3. Neurofibroma

B. Parotid gland
- 1. Cylindroma

C. External ear tumors
- 1. Epidermoid carcinoma
- 2. Basal cell carcinoma
- 3. Melanoma

D. Scalp tumors
E. Nasopharynx
- 1. Carcinoma.

Historical Review

In 1941, Guild first discovered the glomus structure in humans. However, it was in 1942 at the Mount Sinai Hospital in New York that Rosenwasser first performed a radical mastoidectomy on a patient with a bleeding mass, which arose in the middle ear and protruded from the external ear canal. The excised tumor was identical in every characteristic to proven carotid body tumors from the neck. However, the presence of a carotid body-like tumor in the middle ear and mastoid without any demonstrable tumor in the neck had never been described previously. After reading Guild's original description, Otani and Rosenwasser realized that the excised tumor had developed from a glomus jugular structure in the ear.

One of the problems with discovering a new pathologic entity was in providing an appropriate name. In 1945, Otani and Rosenwasser suggested the term "carotid body-like" tumor because of its similar morphology to the carotid body tumors of the neck. The tumor was redefined as glomus jugular tumor by Winship in 1948. In 1949, Lattes and Waltner

reported a more descriptive name, nonchromaffin paraganglioma, for these tumors. Mulligan in 1950 suggested the name chemodectoma, as he thought this best described neoplasms arising from paraganglionic tissue. Gaffney in 1953 thought that these tumors functioned like chemoreceptor organs and thus "should be regarded as a receptoma". Zettergen and Lindström in 1951 referred to these tumors as glomerocytomas. A problem with each of these names is that the underlying function of glomus bodies in the temporal bone is unclear. When this problem is solved, an appropriate name will be found.

Since the first report of glomus tumors in the middle ear and mastoid, each decade has brought new anatomic and surgical discoveries in the classification and treatment of the disease. In the 1940s, tumors involving the middle ear and mastoid that were previously reported as endotheliomas and hemangioendotheliomas were re-evaluated and, in some instances, reclassified as glomus tumors. In the 1950s Rosenwasser, Shambaugh, Dockerty and colleagues, and Le Compte began to collect a series of patients with glomus tumors for indepth study. Surgical removal was limited by the lack of an accurate delineation of the extent of the tumors. Therefore, the focus of treatment was on radiation therapy. By the 1970s, advances in radiographic techniques led to more accurate classification of these tumors. New surgical approaches such as suboccipital, transtemporal, transcochlear, and infratemporal fossa techniques were developed, providing access to all areas of the temporal bone. In the early 1980s, high-resolution computed tomography scanning and new approaches in skull base surgery helped to allow full surgical removal of all types of glomus tumors.

Pathology

Glomus formations similar to those described by Guild were discussed in the anatomic literature in 1840, when Valentin described a slight swelling around the tympanic nerve, which he termed the ganglion tympanicus. In 1878 Krause described the similarity between what he called the glandula tympanica and the glandula carotica.

The original histopathologic description by Otani is still precise today. The general histologic picture is similar to that of a carotid body. The original pathology report was as follows:

... a fragment of tissue covered with a squamous cell layer and immediately beneath this layer is an unusual tumor structure. Large tumor cells in groups are separated by dense fibrous tissue septa which contain dilated vessels. The groups of tumor cells are bordered by capillaries and form small alveolar structures. This alveolar arrangement of the tumor cells is not always distinct and they show compressed cord-like structures in places. The well-preserved tumor cells are large and polyhedral and their nuclei are rather small and not hyperchromatic. The large cytoplasm is frequently vacuolated and this vacuolization may obscure cell outlines. The oval nuclei are uniform, although there are areas in which the tumor cell nuclei vary inshape. There are no mitotic figures.

General Description

Glomus tumors occur in all age groups. Cases have been reported in patients from 16 to 83 years of age, with an average age of 52 years (Ogura et al, 1978). The possibility exists that glomus tumors may arise in the temporal bone at an early age. For example, there is a

report of a child 6 months old having a carotid body tumor (Gosalves and Briant, 1979). Women are more likely than men by a 6:1 ratio to develop these tumors (Spector, 1978). In contrast, in the familial chemodectoma group, the radio between men and women is closer to 1:1 (van Baars et al, 1981). The duration of symptoms before diagnosis ranges from a few weeks to 42 years (Gardner et al, 1977). On the average, symptoms persist from 1 to 3 years before diagnosis (Spector et al, 1975). Reported mortality rates from these lesions range frmo 2 to 22 per cent, and the results from the most recent series fall at the lower end of this range (Rosenwasser, 1973). Cranial nerves are involved in roughly 37 per cent of all cases, and intracranial involvement is manifested in 18 per cent (Bickerstaff and Howell, 1953). The incidence of second glomus tumors is 7 to 8 per cent (Spector et al, 1975).

In rare instances, patients with glomus jugulare tumors may present with all the signs and symptoms of a pheochromocytoma. In 1949, Le Compte first described blood pressure changes in an animal injected with glomus extracts. In 1962, Rosenwasser reported the rare instance of a patient with a glomus jugulare tumor who had severely alarming blood pressure variations during and immediately after surgery. Uncontrollable hypotension resulted, and the patient died within 5 hours after the operation. Since then, the endocrine activity of glomus tumors has been better described. The incidence of functioning endocrine activity in these tumors ranges from 1 to 3 per cent (Schwaber et al, 1984).

Zak and Lawson in 1982 found only 20 reported cases of paragangliomas with endocrine function. In 1980, Farrior reported a case with carcinoid features, including explosive diarrhea, headaches, and facial flushing. These investigators noted that endocrine activity occurred more frequently in males than in females in the group they had reviewed. In 1984, Schwaber and associates reported three cases of endocrine-active lesions: one patient had a dopamine secreting tumor, another had obvious hypertension secondary to the tumor, while the third manifested only subclinical endocrine activity.

Over 160 patients with familial glomus tumors have been reported in the literature, including those with carotid body and glomus vagale tumors (van Baars et al, 1981). Some reports suggest that an autosomal dominant hereditary pattern with incomplete penetrance exists (Parkin, 1981). What is remarkable is the case of 11 or 12 members of a family, spread over multiple generations, who have glomus tumors. In one series of familial glomus tumors, the most common lesion was the carotid body tumor. The most common combination of tumors was glomus jugulare and carotid body.

Multiple tumors occur more commonly in the familial cases. Parkin estimates that the percentage may be as high as 25 to 35 per cent. In contrast to the usual glomus jugulare tumor, the hereditary ones show a male predominance. Few cases of malignant degeneration have been reported.

Because of the high incidence of familial glomus tumors, involvement of other family members should be considered. Since these tumors lie indolent for several years before manifesting symptoms, a workup of immediate relatives, including taking a careful history, a complete physical examination, and additional testing, may detect early lesions in these vulnerable people. Some physicians recommend screening with dynamic angioscintigraphy; however, its low resolution and false positive rate make the value debatable (Laird et al, 1983).

Benign Versus Malignant

One of the most difficult questions to answer is whether glomus tumors are inherently benign or malignant or whether different forms of the tumor exist. Lattes and Waltner (1949) reported a metastasis of a non-chromaffin paraganglioma to the liver, although this report had been questioned. However, in 1958 Rosenwasser reported two cases of metastases from glomus jugulare tumors, and he had seen two additional cases with pulmonary dissemination. In these patients with metastatic spread, the primary glomus tumor had a benign histological appearance. Similar cases of metastatic spread were reported by Taylor and colleagues (1965) and Monroe and Lore (1977). The awareness of the possibility of malignant spread should stimulate a search for metastatic foci.

A problem with some reports of metastatic spread is that glomus type issue can be found in other parts of the body. These areas include the nose, larynx, brain, dura, trachea, and optic chiasm. Another problem is that metastases can be confused with multicentric tumors. It has been reported that this neoplasm is multicentric in 10 per cent of the patients. Willis (1953) believed that in some cases glomus tumors arose from multiple separate foci of tumor cells. However, the cause of the simultaneous development of these tumors in different anatomic sites is unknown. The answers to these problems are still unclear.

Growth Patterns

Glomus tumors are vascular growths. The blood supply originates from the external carotid, internal carotid, and vertebral system. In a comprehensive study, Hesselink and co-workers (1981) and others demonstrated the major arterial pedicles of a series of glomus tumors and found that the major contributors were the caroticotympanic artery and the inferior, anterior, and superior tympanic arteries as well as the stylomastoid and mastoid branches of the occipital artery, the meningeal branch of the ascending pharyngeal artery, and the petrosal branch of the middle meningeal artery. They stated that "since the glomus tissue is located along Jacobson's and Arnold's nerves and since the inferior tympanic artery supplies both of these nerves, it is not surprising that the inferior tympanic artery is the most common arterial feeder to glomus tympanojugular tumors".

However, for many reasons, it is too simplistic to attribute the blood supply of sizable glomus tumors to individual branches. First, there are multiple potential anastomoses between the intracranial and extracranial systems. Second, each artery supplies overlapping areas in the temporal bone, which may result in the tumor receiving blood from several different sources. Third, although intracranial feeders may be insignificant on arteriography, frequently they may limit the effectiveness of embolization procedures and may be a determining factor in resectability or predicting blood loss.

Moret (1982) raises many important questions in an extensive article on the vascular architecture of tympanojugular glomus tumors. First, he divides the tumors into two groups: multiple-compartmented and single-compartmented. Each compartment mimics Spector and colleagues' outline of patterns of invasion (1979). It is Moret's theory that 85 per cent of glomus tumors have multiple compartments. This is significant, as it helps explain the difficulties in surgical resection and embolization. The extensive areas of involvement make the lesions difficult to excise. At the same time, embolization may be complicated by the

intricate arterial supplies to the tumor and the risk of causing cranial nerve paralyses. Moret hypothesizes that "single-compartmented tumors might be unicentric, with a single origin, rising from one group of cells and enlarging by multiplication", whereas multiple-compartmented tumors may be multicentric with simultaneous outbreaks in various locations.

Classification

As a result of advances in neuroradiographic, angiographic, and surgical techniques, classification used to differentiate glomus tumors have changed over the past 40 years. As described by Alford and Guilford in 1962, glomus tumors were traditionally divided into two types: tympanicum and jugulare. This classification system is based on the fact that tumors that develop away from the jugular bulb (glomus tympanicum) have an improved prognosis and are technically less complicated to remove surgically.

In 1969, McCabe and Fletcher divided glomus tumors into three categories based on the degree of bone destruction caused by the tumor. In group I, there is no bone destruction, the facial nerve is intact and the jugular foramen is unaffected. Characteristically, the lesion is a glomus tympanicum tumor that arises on the promontory or the floor of the tympanum. In group II, the tumor extends beyond the middle ear to involve the aditus, antrum, or mastoid and the jugular bulb. Bone destruction is limited to the mastoid, and the facial nerve may be paretic. In group III, there is widespread extension of the tumor intracranially or to the base of the skull. Bone destruction involves the petrous bone, jugular fossa, and occipital bone, and the patient may have the jugular foramen syndrome.

Jenkins and Fisch in 1981 redefined the classification of glomus tumors to take into account improved surgical planning. Their classification system involves four groupings: A, B, C, and D. Type A includes tumors of the middle ear. Type B includes tumors involving the mastoid area without infralabyrinthine extension. Type C includes tumors extending into the infralabyrinthine area and the petrous apex. Type D includes intracranial extension and contains three subgroups. Type D1 represents tumors with intracranial extension "less than 2 inches in diameter". Type D2 includ tumors with greater than 2 inches extension, whereas Type D3 covers tumors presenting inoperable intradural and intracranial extension. An alternative classification was proposed by Jackson and colleagues in 1982.

Diagnosis

Tumors that enlarge concentrically and erode the bony floor of the middle ear cavity may invade the cochlea and compress or invade the dome of the jugular bulb. Symptoms produced by a glomus tumor are related to the vascular nature of the lesion and its propensity to erode surrounding structures. Unilateral pulsatile tinnitus may be the patient's initial complaint. As the tumor enlarges, it can destroy the ossicles, involve the facial nerve, and grow through the drum, presenting as a vascular pulsatile polyp. Erosion into the cochlea will produce a neurosensory hearing loss and vertigo. Involvement in the base of the skull near the jugular foramen may cause paresis of cranial nerves IX, X, XI or XII. Intracranial spread may produce symptoms that mimic a brain tumor.

Clinically, patients with glomus tumors may present with many different signs and symptoms. For example, a conductive hearing loss, a slight bulging tympanic membrane, and

mild tinnitus in a patient may signify serous otitis media as well as a glomus tumor. Similarly, a patient with a bleeding, buzzing, and bluish eardrum may have chronic cholesterol granulomatous mastoiditis, as opposed to a glomus tumor. In reviewing the major series of glomus tumors over the last 25 years, one finds that the most common symptoms, in order of frequency, are hearing loss, tinnitus, discharge, otalgia, vertigo, bleeding, cranial nerve palsies, a mass in the external canal, a mass visible behind the eardrum, and a positive Brown's sign. If the tumor extends into the base of the skull or erodes bone over the facial canal, then the patient will present with cranial nerve abnormalities. The most commonly involved nerves, again in order of frequency, are the facial (VII), vagus (X), hypoglossal (XII), glossopharyngeal (IX), and spinal accessory (XI).

Traditionally, routine x-ray studies of the middle ear, mastoid, and temporal bone were used to aid in the diagnosis of glomus tumors. In addition, polytomography was used to delineate bone destruction around the jugular bulb and carotid artery. During the 1970s, computed tomography (CT) replaced these methods as the primary diagnostic tool for glomus tumors of the temporal bone. Combined with angiography, CT scanning with contrast helps to classify and outline lesions with respect to both intratemporal versus extratemporal and intracranial versus extracranial tumor growth. Preoperative evaluation and postoperative treatment can thus be refined based on high-resolution CT scans (Wright et al, 1979).

Radiographically, glomus tumors involve the temporal bone in several common patterns:

1. The tumor may spread from the jugular bulb area through its bony covering and extend into the middle ear. It may displace or destroy the ossicles and erode the facial nerve or the labyrinth. Extension into the mastoid air cells may also occur.

2. The tumor may arise at the site of the jugular bulb and grow anteroinferiorly, exposing the carotid artery and extending into the petrous apex.

3. The tumor may gro from the jugular bulb medially, eroding the hypoglossal canal and the occipital condyle and extending into the posterior fossa.

4. The tumor may occasionally arise from glomus bodies associated with the vagus nerve. In such cases, the main tumor mass may be located endocranially and may secondarily involve the temporal bone.

Angiography with subtraction techniques represents an important supplement to CT scans in the diagnosis and treatment of glomus tumors. First, angiography allows the surgeon to identify the various arterial supplies to the tumor. Second, it helps to delineate extension into the jugular vein and carotid artery. Third, it enables the diagnosis of multiple or bilateral lesions in cases where only one lesion has demonstrated symptoms. Finally, it aids in the differentiation of arterial and venous malformations from glomus tumors (Sinnreich et al, 1984; Tjellstram and Svendsen, 1983).

Other types of vessel studies may also be used in select cases or where the needed expertise is routinely available. Retrograde jugulography may be used to see if the lumen of the vein is blocked. Digital subtraction antiography is useful when a patient cannot tolerate

large amounts of contrasts or long periods of catheterization. Radionuclide angiography has been utilized for screening but is severely limited in its clarity.

One of the most exciting areas in radiology today is therapeutic embolization. Hilal and Michelsen first reported the use of embolization of neoplasms of the head and neck in 1970. Gelfoal, liquid Silastic, radiopaque Silastic spheres and Teflon particles (Ivalon) have all been used as embolization particles. The primary purpose of embolization is to reduce tumor vascularity and thereby reduce blood loss intraoperatively. Embolization has been used preoperatively and for palliation in unresectable glomus tumors and in patients who could not tolerate surgery. Additionally, its advantages include the fact that it is repeatable, can be performed under local anesthesia, and may relieve some symptoms. However, it has several limitations: multiple anastomoses may restrict its benefit, occluded vessels may recannulate, and neurologic complications may result.

Treatment

Choice of Therapy

Clinical experience with glomus tumors has indicated that the treatment of choice is complete surgical excision. Irradiation may be appropriate therapy for advanced lesions that involve the petrous apex or for thos that extend intracranially and therefore cannot safely be totally excised without extensive mutilating surgery. However, it has become apparent that no consensus exists regarding the choice of therapy, since a lesion that one physician may consider too advanced for surgery may be successfully operated on by another. Furthermore, it is difficult to determine the efficacy of the different types of therapy recommended for glomus jugulare tumors.

Since it is known that without treatment these neoplasms may not change significantly over the years, the assessment of results achieved by surgery, irradiation, or a combination of these two modalities is difficult. Accordingly, the decision as to the ideal form of therapy must be individualized. The physician must consider the patient's age and general health, as well as the location and size of the tumor. Thus, when an octogenarian presents solely with a hearing loss caused by a large glomus tumor involving the base of skull, the clinician has several therapeutic options. In view of the patient's advanced age and restricted life expectancy, the surgeon may elect to defer any treatment until additional symptoms appear that necessitate palliation. Alternately, irradiation may be recommended. In contrast, if a healthy, younger person presents with a similar lesion, it may be feasible to attempt resection, which may be followed by postoperative irradiation.

Surgery

The variety of procedures available to the surgeon has recently expanded dramatically. Many of the unsolved problems regarding control of bleeding, exposure to the skull base, and control of internal carotid, sigmoid sinus, and intracranial extension have been addressed with some success. Smaller lesions limited to the hypotympanum or mastoid are easily accessible through transcanal or transmastoid approaches. However, exposure to the skull base necessitates the skills of the head and neck surgeon. Similarly, intracranial spread can be approached only by the specially trained neurotologist, often in concert with a neurosurgeon.

Equally important are the questions of surgical staging and the determination of the limits of resection. Patients with multiple glomus tumors may require multiple separate procedures, particularly if the tumors are on contralateral sides of the head and neck. The treatment of lesions with intracranial involvement is controversial. There is debate as to whether the otologist should perform the temporal bone and neck exploration at the same time that the neurosurgeon approaches intracranial extension through a suboccipital approach to the posterior fossa, or whether the two procedures should be staged separately. With the advent of transoral approaches to the clivus, transsphenoidal approach to the petrous apex, and an infratemporal fossa approach to the skull base, limits of resectability are rapidly expanding. Jackson (1982) emphasized that his "surgical criteria for unresectability remain extensive foramen magnum extension, disease into the cavernous sinus, and the medical infirmity usually associated with age".

The surgical approach depends on the limits of the tumor, its classification, and the abilities of the surgeon. Glomus tympanicum tumors are resected in one of three ways: a middle ear exploration, mastoidectomy, or the hypotympanic approach of Farrior (1984). The type of mastoidectomy may be either a canal wall up procedure with an extended facial recess approach or canal wall down procedure. The key to preservation of the facial nerve without avulsion of the stapes is positive identification of these structures before the tumor removal is begun, while the operative field is relatively free of bleeding. Experience has shown that the most efficient technique for resecting these vascular lesions expeditiously is removal of all bone from around the tumor, with care being taken not to manipulate the mass before complete exposure has been achieved. During tumor resection, the troublesome bleeding is controlled by pressure packing. After removal, the feeding arterial vessels can be individually identified and microcauterized.

Glomus jugulare tumors frequently need more extensive surgery, because of the involvement of wider areas along the temporal bone and the base of the skull. Useful techniques are the infratemporal fossa approach of Fisch (1978), combined operations with neurosurgery as well as other base-of-skull operations. By using these procedures, it is possible to gain access to the parasellar area, the clivus, eustachian tube, petrous apex, and base of the skull. Several authors, including Glasscock, Jackson, Gardner, and Spector, have written on this subject and made various modifications. Each of these approaches is balanced against the procedure's complications, including malocclusion, multiple cranial nerve paralyses, intracranial problems, and hearing loss (Glasscock et al, 1978; Gardner et al, 1981; Spector and Sobol, 1980).

Irradiation

The inherent tendency of glomus jugulare tumors to grow slowly may be more important in the determination of their radiocurability than is their responsiveness to irradiation. If one employs irradiation in the treatment of those tumors that are not thought to be surgically removable, the best results can be obtained when the limits of the tumor are precisely defined. This enables the radiotherapist to substantially reduce the volume of normal tissue that must be exposed to radiation while simultaneously ensuring that the entire tumor is properly treated.

Surprisingly, there is still wide disparity of opinion as to the amount of radiation necessary to cure these tumors. Treatment with 3000 to 6000 rads over a period of 3 to 6 weeks has been recommended. Others have recommended various combinations of therapies, usually under 5000 rads (Wang, 1980).

In view of the unpredictable growth rate of glomus jugulare tumors, one cannot be dogmatic in regard to the effectiveness of irradiation. Bickerstaff and Howell (1953) reported a glomus jugulare tumor involving the glossopharyngeal, vagus, spinal accessory, and hypoglossal nerves in a person who was not treated and who died 42 years after the onset of symptoms. One should speak of years of survival rather than radiocurability when discussing results of treatment. These tumors have been regarded as radioresistant, but this does not mean that irradiation is useless. Frequently, response to irradiation may be slow, and the tumor may continue to shrink for months after treatment.

Morbidity and Mortality

Patients with glomus tumors have a small but real incidence of mortality resulting from treatment. In 1952, Winship and associates collected 62 cases from the literature and found the mortality rate to be 21.5 per cent. In Spector and Sobol's 1980 series of 77 patients, there were seven deaths resulting from either the tumor or the surgery. In addition, there were six deaths from other malignancies and four from other causes. In Fisch's 1978 series of 51 patients needing infratemporal fossa surgery, there was only a 2 per cent mortality rate, whereas in Jackson and colleagues' 1982 series of 70 patients there were no deaths. The mortality rate has clearly decreased in recent years as the result of earlier detection, better surgical exposure, and a defined approach to minimize bleeding. However, these lower mortality rates occur only in the hands of those physicians trained to handle the myriad of intraoperative and perioperative problems.

Each year, as the limits to surgery expand and the degree of perioperative morbidity slowly increases, the most common surgical complication is facial nerve paresis or paralysis. Also, difficulty in swallowing and aspiration often trouble patients after skull base surgery, frequently owing to manipulation of the nerves. In Fisch's 1978 series, 39 per cent of the patients showed facial nerve changes; in Spector and Sobol's 1980 series, the incidence of nerve change was only 19 per cent. In Jackson and co-workers' 1982 series, postoperative complications included paralysis of cranial nerve IX in 18 per cent of patients, of cranial nerve X in 27 per cent, of cranial nerve XI in 9 per cent, and of cranial nerve XII in 9 per cent. Therefore, care must be taken to make the appropriate diagnosis and to be prepared for secondary complications, including aspiration and dysphagia, should they arise (Cece et al, 1987).

Neurogenic Tumors of the Temporal Bone

Schwannomas (Neurilemmomas)

The principal neurogenic tumors of the cranial nerves entering and exiting the temporal bone have a common origin in the Schwann cells of the nerve sheath. The nomenclature used in the classification of these tumors has been inconsistent. The terms schwannoma, neurilemmoma, neurinoma, and (in the case of cranial nerves VII and VIII,

respectively) facial nerve neuromas and acoustic neuromas are all synonyms for a benign, encapsulated, slowly growing nerve sheath tumor that can arise from any cranial nerve with the exception of the olfactory and optic nerves.

Schwannomas (neurilemmomas) are characterized histologically by a distinct capsule, so-called Verocay bodies (pallisades of nuclei), and the Antoni A and B growth patterns. Mitoses are sparse to absent, and these tumors rarely if ever undergo malignant change. Schwannomas are the most common of the neurogenic tumors of the temporal bone. They may occur on cranial nerve VIII in the internal auditory canal or cerebellar pontine angle, and in this location they are more commonly referred to as acoustic neuromas. Schwannomas may also occur on cranial nerve VII intratemporally, and in this location they are more commonly termed facial nerve neuromas. Clemis (1980) has provided an excellent review of the classification of schwannomas.

Neurofibromas

Neurofibromas are less common than schwannomas. In distinction to schwannomas, neurofibromas lack a capsule and also lack the Antoni A and B growth patterns so characteristic of schwannomas. Histologically, neurofibromas are composed of proliferating axons and Schwann cells with undulating cell processes, in a matrix that varies from fibrous to myxoid and usually contains numerous mast cells (Barnes et al, 1985). The diagnosis of solitary neurofibroma is always conditional, as it may herald the onset of diffuse disease and may be the first sign, especially in young individuals, of von Recklinghausen's disease.

Von Recklinghausen's disease (multiple neurofibromatosis) is a systemic disorder with autosomal dominant inheritance. The disease is characterized by multiple neurofibromas of the skin and internal structures, café au lait spots, and mesenchymal dysplasia of the blood vessels and bones.

Central, peripheral, and visceral forms are present, as well as incomplete expressions of the disorder. Von Recklinghausen's disease is generally evident at an early age, and the prognosis depends on the site of involvement. Central forms are associated with multiple intracranial manifestations including heterotopic gray matter within cerebral white matter, causing distortion of the cerebral architecture (Kimmelman, 1979). Other intracranial abnormalities include meningiomas, which may be multiple, and schwannomas, particularly of cranial nerve VIII. When schwannomas of cranial nerve VIII occur in multiple neurofibromatosis, they are generally bilateral, and they may be the principal or the only manifestation of von Recklinghausen's disease (Nager, 1964).

Other sites of neurogenic tumors within the temporal bone include the jugular foramen and the petrous apex. Gacek (1976, 1983) has reported on neurofibromas of the jugular foramen. Clinically, these lesions are characterized by pulsatile tinnitus, hearing loss, a middle ear mass, and involvement of the nerves of the jugular foramen, IX, X, XI, and XII. In addition, jugular foramen neurofibromas can extend intracranially and can also involve cranial nerves VII and VIII. Petrous apex neurofibromas may occur. These lesions arise in the neural elements in the foramen lacerum and include the greater superficial petrosal nerve and the deep petrosal nerves. Symptoms are predominantly those affecting the trigeminal nerve. Rarely, other sites in the temporal bone may be involved, including intralabyrinthine lesions

that are generally appreciated only post mortem on temporal bone examination (Johnson and Kingsley, 1981).

Facial Nerve Neuromas

Commonly referred to as facial nerve neuromas, primary tumors of the facial nerve originate from the Schwann cells of the nerve sheath. Two types of tumors of Schwann cell origin can occur in the facial nerve: neurilemmomas and, very rarely, neurofibromas. These tumors may be difficult to distinguish from one another in small specimens. Primary facial nerve tumors are uncommon, with 140 cases reported in the world literature; however, their incidence may be higher. Saito and Baxter (1972) observed five previously asymptomatic neurilemmomas in a review of 600 temporal bones, giving an upward incidence of 0.8 per cent.

The symptoms produced by a facial nerve neuroma will be related to its location. Within the temporal bone, facial paralysis is the most frequent presenting symptom. Janecka and Conley (1985) reported that facial nerve paralysis or weakness was presented in 84 per cent of 17 patients with intratemporal facial neuromas. In the same publication they also demonstrated the relative rarity of neurofibromas versus neurilemmomas. Fisch and Ruttner (1977) emphasized that seven of nine intratemporal facial nerve neuromas in their series arose from the region of the geniculate ganglion. According to these investigators, embryologically the facial nerve undergoes structural reorganization at the geniculate ganglion, and this is the reason for the predilection for this site of neoplastic and hamartomatous lesions.

Classically, facial paralysis of neoplastic origin is gradual in onset and slowly progressive. However, facial paralysis of sudden onset also can be the result of neoplasm. Other signs of facial nerve neuromas include fluctuating paresis, with incomplete return of facial nerve function, as well as hemifacial spasms. Facial nerve neuromas cause a gradual compression of the nerve as the benign tumor enlarges rather than neoplastic destruction. Unresolved "Bell's" palsy of 4 to 6 months' duration must be investigated to rule out neoplasms.

The second most frequent symptom of facial nerve neuromas is hearing loss. The hearing loss may be the result of conductive, cochlear, or retrocochlear involvement. When the neuroma arises in the vertical portion, a facial paralysis will be present and the hearing may be unaffected. However, when the neuroma arises in the horizontal portion, the ossicles may be affected, producing a conductive hearing loss. If the neuroma arises in the area of the geniculate ganglion, infiltration of the labyrinthine structures may produce a sensory hearing loss and may also affect vestibular function. A vestibular nerve schwannoma may occur simultaneously with a facial neuroma, as described recently in two cases by Jung and colleagues (1985).

When a facial nerve neuroma is suspected, a complete neurotologic examination should be performed. Complete audiologic evaluation is obtained as well as electronystagmography in those patients with sensorineural hearing loss or vestibular symptoms. Polytomographic radiologic studies to visualize the intratemporal neuromas are being rapidly supplanted by the use of high-resolution CT scanning. High-resolution CT scanning with intravenous contrast has the advantage of providing visualization of the cerebral

potine angle and better visualization of the cerebral pontine angle and better visualization of the geniculate ganglion region and horizontal segment of the facial nerve (Latack et al, 1983).

Contemporary surgical management of these lesions is based on three regions of presentation: intratemporal, intratemporal with extratemporal extension, and intratemporal with intracranial extension. Surgical approaches to these regions have been reviewed by Pulec (1972), Horn and co-workers (1981), Bailey and Graham (1983), and Pillsbury and associates (1983). Treatment for facial nerve neuromas involves surgical resection with primary nerve grafting, or rerouting of the facial nerve with primary approximation. Intratemporal lesions with intracranial extension are treated by combined intracranial and extracranial approaches. When the neuroma involves the intralabyrinthine segment of the facial nerve or the geniculate ganglion, a middle fossa approach is required. Hypoglossal-facial anastomosis may be necessary for rehabilitation if the proximal nerve cannot be sutured.

Osseous Lesions

Osteomas and Exostoses

Osteomas may occur anywhere in the mastoid or temporal bone but are most frequently clinically significant when they arise in the external auditory canal. Osteomas are to be distinguished from exostoses. Both lesions remain asymptomatic, unless because of increasing size they result in obstruction of the external auditory canal. As defined by Sheehy (1958), osteomas are grossly pedunculated, unilateral, solitary masses covered by squamous epithelium of the external auditory canal. They appear to arise from the tympanosquamous suture line, lateral to the isthmus, and less commonly from the tympanomastoid suture line. It is unknown why osteomas occur along the suture lines, although the bone is more vascular than the overlying skin and subcutaneous tissue is thicker than along other surfaces of the tympanic bone.

Exostoses, on the other hand, are broad-based, bony growths of periosteal origin, are generally multiple and bilateral, and can vary in size from tiny nodules to masses obstructing the external auditory canal. Exostoses are distinctively more common in cold water swimmers and surfers. Fowler and Osmun (1942) demonstrated in a guinea pig model that repeated irrigation of the external auditory canal resulted in the diffuse production of new bone in the middle ear, adjacent to the area of irrigation. Harrison (1951) postulated that prolonged vasodilation, which followed the presence of cold water in the deep meatus, was responsible for the development of exostoses. DiBartolomeo (1979), in a complete review of the subject of exostoses, concluded that irritation, from whatever source but particularly thermal irritation, resulted in thickening of the tympanic ring and the development of exostoses.

Graham (1979), utilizing scanning electron microscopic techniques, demonstrated clearly that osteomas are characterized by an abundance of fibrovascular channels throughout, surrounded by lamellar bone oriented in multiple directions. In contrast, the internal structure of exostoses is characterized by dense, parallel, concentric layers of subperiosteal bone, abundant in osteocytes and devoid of fibrovascular channels.

Exostoses or osteomas can become clinically significant when their size results in the retention of cerumen and of debris medial to them. Subsequent recurrent episodes of external

otitis media accompanied by conductive hearing loss are indications for surgical removal. Exostoses are considerably more common than osteomas by a ratio of 5 to 1. Lesions of either type requiring surgical treatment are relatively uncommon. In a review of 80 cases requiring surgical treatment, Sheehy (1982) concluded that osteomas could most frequently be removed transcanal, but exostoses were best managed by a postauricular approach when surgery was necessary.

Extracanalicular osteomas of the temporal bone are uncommon. The lateral cortex of the mastoid is the most frequent site of occurrence. Very rarely, osteomas have been described within the middle ear and the eustachian tube. If a decision is made to remove an osteoma of the mastoid because of increasing size and deformity, a cleavage plane can generally be encountered between the tumor and normal bone of the external cortex. Mastoid osteomas rarely may extend into the fallopian canal and bony labyrinth, and in these cases complete removal may not be indicated (Denia et al, 1979).

Giant Cell Tumors (Osteoclastomas)

Giant cell tumors (osteoclastomas) of bone represent 4 to 5 per cent of all primary neoplasms of bone. Only approximately 2 per cent of giant cell tumors, however, occur in the head and neck region. Most frequently, giant cell tumors involve the long bones in the epiphyseal region. In the skull, giant cell tumors occur in the sphenoid, ethmoid, and temporal bones. These bones are of endochondral origin, whereas the remainder of the cranial bones with the exception of the occipital bone are of membranous origin. With less than 25 cases of giant cell tumor of the temporal bone reported in the world literature, this neoplasm is a true rarity.

Hearing loss, pain, and cranial nerve paralysis, generally of the facial nerve, are the most likely presenting complaints. Patients with giant cell tumors of the middle ear and mastoid may present with a middle ear mass and a conductive hearing loss, whereas patients with petrous involvement may present with a sensorineural hearing loss and vertigo. In patients with large lesions of the petrous apex, there may be contiguous involvement of the sphenoid and symptoms of headache and diplopia.

Although these lesions are considered benign neoplasms, they may behave aggressively, and their counterparts in the long bones may metastasize. Local recurrence rates for giant cell tumors are high, and they may occur in relatively inaccessible regions of the skull base.

Grossly, giant cell tumors are reddish brown in appearance due to prior hemorrhage within them. Histologically, they are characterized by multinucleated giant cells of the osteoclastic type, in a stroma of ovoid or spindle shaped cells. The giant cells are thought to arise from the fusion of stromal cells. In small biopsy specimens, giant cell tumors may be histologically difficult to distinguish from giant cell granulomas. In order to differentiate these lesions from one another, the age of the patient, site of the lesion, and clinical course are important.

Rosenwasser (1969) reported on a case in a 51-year-old man who complained of hearing loss of 10 months' duration. Facial weakness was noted, and a mass was present in

the posterosuperior quadrant of the middle ear behind an intact tympanic membrane. Mastoid x-rays reveal destruction of the tegmen with diffuse clouding. At operation, the middle ear was filled with tumor, and the facial nerve was exposed in its tympanic segment. Middle fossa tegmen was absent in the region of the geniculate ganglion. The defects in the tegmen extended anteriorly toward the eustachian tube.

Clinically, the lesion felt firm and friable. It was reddish in color and resembled organized granulation tissue. It did not have a capsule. The resected tumor fragments showed numerous giant cells, osteoclastic type, and so-called stroma cells in the background; the latter showed swollen fibroblast-like cells, but occasionally there were elongated cells with double or triple nuclei, which suggested that they were precursors of giant cells. There was considerable iron pigment scattered throughout, indicating old hemorrhage. Areas of fresh hemorrhage were also present. A few fragments of tissue were covered by squamous cell epithelium. The expanding giant cell tumor, destroying bone cortex, had invaded the soft tissue and was extending underneath the epidermis.

Perhaps because of their rarity and similar histology, considerable confusion exists regarding the appropriate classification of giant cell lesions. In an excellent article, Smith and Ward (1978) reviewed giant cell lesions of the facial skeleton and differentiated three distinct types: brown tumors of primary and secondary hyperparathyroidism, giant cell granulomas, and giant cell tumors (osteoclastomas). Brown tumors of primary and secondary hyperparathyroidism can be distinguished by appropriate metabolic, endocrine, and renal evaluation. Giant cell granulomas can occur peripherally, most commonly in the gingiva and palate. Centrally, giant cell granulomas occur in intraosseous locations, generally within the alveolus. Giant cell granulomas occur in children and young adults. Histologically, their stroma is more fibrous than that of giant cell tumor and is largely devoid of collagen. Treated surgically with adequate curettage of the involved bone, giant cell granulomas are generally cured.

Giant cell tumors, on the other hand, are more common in middle-aged and elderly patients. They are locally destructive and aggressive. Local curettage alone may not be adequate in a significant number of cases, and recurrence locally is relatively high. Radiotherapy has been employed for giant cell tumors involving the skull base; however, there is a possibility of secondary malignant degeneration. Combined intra- and extracranial approaches, such as that described by Hunt and Glasscock (1974), may be necessary to eradicate extensive lesions of the temporal bone.

So-called primary malignant giant cell tumors have been described, in which areas of giant cells and stroma typical of giant cell tumors coexist in the same specimen, with areas histologically typical of osteosarcoma, fibrosarcoma, or fibrous histiocytoma (Barnes and Verbin, 1983). According to Rosenwasser, these lesions would be better described only in reference to their malignant component and cannot properly be termed giant cell tumors.

Fibrous Dysplasia

Fibrous dysplasia is a benign disease of bone characterized by the abnormal proliferation of fibro-osseous tissue within cancellous bone. The lesion displaces normal cortical bone by erosion and expansion from within; the cortex may become very thin but

remains intact. Fibrous dysplasia represents approximately 7 per cent benign bony tumors; however, its occurrence in the temporal bone is rare. Nager and colleagues (1982) presented a detailed monograph on the subject and found 69 cases of involvement of the temporal bone in the world literature. The mean age at presentation in these investigators' series was 28. Females were more commonly affected than males by a ratio of 2:1.

Two forms of fibrous dysplasia are recognized: (1) monostotic (unifocal) form and (2) a polyostotic multifocal form. The polyostotic form is less common and represents approximately 20 to 30 per cent of cases. The bony lesions in the polyostotic form are multiple and characteristically unilateral in both the cranial and long bone presentations. The polyostotic form may occur in conjunction with abnormal mucosal pigmentation and precocious puberty, in which case it is termed the McCune-Albright syndrome.

Clinical symptoms most commonly referable to temporal bone lesions are due to progressive bony occlusion of the external auditory canal by the expanding bony lesion. This occlusion leads to conductive hearing loss as well as to complication secondary to the entrapment of cerumen and squamous epithelial debris medial to the obstruction. Canal cholesteatomas also can occur from the bony stenosis. Expansion of the bony lesions in other locations within the temporal bone may lead to intracranial mass effects, and in the lateral mastoid and root of the zygoma will produce progressive swelling and cosemtic deformity.

Fibrous dysplasia is a locally circumscribed lesion that on gross examination has a soft gritty texture and may appear red to white in color. Histologically, the lesion is characterized by irregular trabeculae of immature woven bone, in distinction to normal lamellar bone, without functional oritentation and in a stroma of collagen matrix made up of cellular fibrovascular connective tissue. The classic radiologic appearance of fibrous dysplasia has been described as that of a cyst-like lucency with a ground glass appearance on plain films. This classic pattern was categorized by Fries (1957) into three distinct types: pagetoid, sclerotic, or cystlike. Pagetoid lesions have the appearance of alternating areas of sclerosis and lucency, accompanied by widening of the inner and outer tables of the calvarium, producing an appearance similar to Paget's disease (osteitis deformans). Sclerotic lesions are homogeneously dense; cystlike lesions are spherical or ovoid, lucent, and have a dense boundary. CT scanning has added to the ability to determine the extent of lesions, especially those that involve the base of the skull. Within the temporal bone, the expansile lesion may have a cystlike homogeneous appearance. The otic capsule, however, is spared, and the labyrinth has the appearance of floating within the lesion (Bergeron and Osborne, 1984).

Ossifying fibromas are similar lesions but can be differentiated from fibrous dysplasia by clinical, radiologic, and histologic parameters. Ossifying fibromas are very rare lesions that primarily affect the mandible and maxilla. Only several cases within the temporal bone have been documented (Stecker, 1971; Levine et al, 1981). Histologically, ossifying fibroma differs from fibrous dysplasia in that the fibrous component of the fibro-osseous process predominates. Trabeculae are made up of centers of woven bone, surrounded by lamellar bone rimmed by osteoblasts; in contrast, in fibrous dysplasia osteoblasts are absent or rare. Radiologically, ossifying fibromas are considered to have more distinct margins, which may be sclerotic.

Treatment of both lesions is surgical when the lesions become clinically significant. Radiotherapy is contraindicated because of the possibility of malignant transformation. Sarcomas can arise within fibrous dysplasia, and their occurrence is estimated to be 0.5 per cent. Huvos and colleagues (1972) reviewed 12 cases of carcomas arising in fibrous dysplasia lesions. Only one of these cases had prior radiotherapy, and the other lesions had arisen de novo within fibrous dysplasia lesions. These investigators emphasized that primary bone sarcomas appear more frequently in patients whose skeletal system is affected by other benign conditions. As far as we know, no cases of sarcoma of the temporal bone arising in fibrous dysplasia lesions have been reported to date.

When fibrous dysplasia produces significant clinical symptoms, surgery is indicated. Indications include bony encroachment of the external auditory canal, which causes sufficient narrowing to produce a conductive hearing loss, recurrent infection, or secondary canal cholesteatoma. Restenosis can be a problem unless a very wide bony canal is created at the time of surgery. In our cases, we have utilized the immediate relining of the wide bony canal created at surgery with a thin split thickness graft. These grafts prevent formation of granulation tissue, provide for rapid healing, and prevent contracture of soft tissue. We have not found it necessary to use Silastic stents to maintain patency of the bony canal following surgery, although this has been advocated by Nager and associates (1982).

Choristoma of the Middle Ear

Choristomas (aberrant salivary glands) have been reported in a number of areas of the head and neck, but only 12 cases have been reported in the ear. Despite its rarity in the ear, salivary gland choristomas arise in conjunction with facial nerve and ossicular chain anomalies.

The first case report of this entity was by Taylor and Marin in 1961. They presented the case of a 31-year-old woman with unilateral conductive hearing loss with a soft mass under the tympanic membrane, adjacent to the facial nerve. The mass was removed surgically, but a temporary facial weakness resulted. During surgery, an inferiorly displaced incus with erosion of the long process was noted. In 1962 Steffan and House reported a 51-year-old woman, with a similar hearing loss and middle ear mass, who had no incus or stapes and whose facial nerve was surrounded by the mass in the horizontal portion. Another abnormality reported is the absence of the ossicular muscles, including the stapedius and tensor tympani. Peron and Schknecht in 1975 reported a 20-year-old patient with bilateral salivary gland choristomas in the middle ear. This patient also had inner ear abnormalities as well as mental retardation.

In 1984, Kartush and Graham reviewed all previously reported cases. They found the median age of patients to be 22 years, with the range from 6 to 52 years. Females were predominant by a ratio of 2:1. The incus was absent in four cases and malformed in seven. The stapes was absent in five cases, malformed in four, and obscured by tumor in two. In these cases, the facial nerve was vulnerable to injury, since it was dehiscent in seven, displaced in three, and obscured by tumor in two. Other associated abnormalities included abnormal auricles in two cases, supra-auricular masses in three, obliterated oval window in two, and a persistent stapedial artery in one. The combination of ossicular abnormalities and facial nerve problems may be explained by second branchial arch developmental abnormality.

Treatment of middle ear choristoma is conservative surgical removal. In most reported cases, operative intervention was associated with a facial nerve injury. Therefore, incomplete resection of the lesion to preserve the integrity of the facial nerve is justified. No cases involved malignant degeneration or continued growth.

Lipomas of the Temporal Bone

A lipoma is an encapsulated tumor of fatty density usually occurring in the subcutaneous tissue in adults. It is one of the most common tumors in humans. Its occurrence in the head and neck is uncommon, although it may occur in the oral cavity and palate. Lipomas are among the rarest tumors of the temporal bone. To date, six cases involving internal auditory canal and one involving the middle ear have been documented.

Intracranial lipomas have been reported, but they usually occur in the midline near the corpus callosum. In 1978, Olson and colleagues reported two cases of lipomas of the internal auditory canal and cerebellar pontine angle. The two patients presented with long histories of Ménière's symptoms. Polytomography and posterior fossa myelography demonstrated the lesions. In one patient, the tumor was removed via the middle fossa approach; in the other, via a translabyrinthine method. In 1986, the authors treated a 4-year-old girl with multiple develomental defects who had an isolated lipoma of the middle ear (Selesnick et al, 1990). No facial nerve or ossicular problems were noted.

Lipomas are rare in children, although congenitaland familial forms do exist. Multiple classifications for fatty tumors exist and are often a source of diagnostic confusion. In pediatric cases, often it is unclear whether these are true neoplasms, since the distinction between discrete masses and hyperplasia may be unclear. The origin of fatty tissue tumors is debatable; some speculate that it is from pluripotential embryonic mesenchyme, whereas others believes that it is due to metaplasia of connective tissue or hyperplasia of normal fat cells.

Squamous Cell Carcinoma

Squamous cell carcinoma of the external ear canal, middle ear, and temporal bone is not a common disease; however, it is the most common malignancy of this region. Since the early symptoms of squamous cell carcinoma of the ear may masquerade as chronic otitis externa or chronic otitis media, the correct diagnosis is frequently made after the disease has spread to vital structures within or beyond the confines of the temporal bone. Accurate comparison between one clinical series and another has been handicapped by the lack of an acurate preoperative staging system for the disease. In addition, the local site of origin of the tumor is frequently unclear. Thus, there may be considerable difficulty in distinguishing whether a lesion has begun in the middle ear, or vice versa. To complicate matters further, preoperative staging in general has grossly underestimated the extent of tumor spread at the time of presentation (Goodwin and Jesse, 1980; Wagenfeld et al, 1980).

The etiology of squamous cell carcinoma of the external ear canal, middle ear, and temporal bone is unknown. A distinct relationship seems to exist between long-standing chronic suppurative otitis media of many years' duration and the development of the disease. This relationship has been demonstrated by many authors over the years and is present in

more than half of the cases reviewd in most large series (Conley and Novack, 1960; Lewis, 1975). The use of tobacco or tobacco products has not been discussed in any reports to date. A cluster of cases was reported in which a direct relationship existed between the exposure to radium paint and the development of the disease (Beal et al, 1965). Prior radiotherapy is believed to have had a direct etiologic role in several reported cases (Ruben et al, 1965). Most authors agree that chronic otorrhea plays a role in the development of the disease, and several published series have demonstrated the simultaneous presence of cholesteatomas in up to 25 per cent of their cases (Lewis, 1973; Coachman, 1951). Michaels and Wells (1980), however, have critically reviewed the histologic and temporal bone materials from 28 cases of squamous cell carcinoma of the middle ear. These authors noted areas of in situ mucosal disease alongside areas of invasive disease, much like squamous cell carcinoma in other sites of the upper aerodigestive tract. None of the patients in their series had concomitant cholesteatomas. They concluded that progressive growth of the tumor results in secondary infection of the middle ear via the eustachian tube and external auditory canal, and that the chronic otorrhea is more likely a result than a cause of squamous cell carcinoma of the ear.

Squamous cell carcinoma of the external ear canal, according to Lewis (1983), is twice as common in females as in males. However, squamous cell carcinoma of the middle ear and mastoid appears to have an equal sex distribution. Barnes and Peel (1985) reviewed the major reported series of the disease and found a peak age incidence to occur in the mid-fifties. The local site of origin may be difficult to assess because of the advanced nature of the lesion. Clinically, however, early lesions may be separated broadly into those that occur in the external ear canal and those that occur in the middle ear. Lesions of the external ear canal may then be subdivided into predominantly membranous or cartilaginous external canal lesions and those of the osseous external auditory canal (Goodwin and Jesse, 1980).

Squamous cell tumors of the external ear canal infiltrate medially. Adams and colleagues (1971) and Crabtree and associates (1975) believe that the majority of middle ear lesions are secondary to the direct extension of lesions of the external ear canal. However, Hyams (1976), Conley and Schuller (1976), and Lewis (1983) have reported a considerable number of cases that they believe were primary in the middle ear. The ratio from Conley and Schuller's series favor the occurrence of external ear canal lesions in a ratio of 3:1.

Regardless of the origin, once the squamous cell tumor has involved the middle ear it then spreads to sites within and beyond the temporal bone. The tumor may extend through the tegmen of the epitympanum and directly involve the dura of the middle fossa (Rosenwasser, 1940). Michaels and Wells (1980) have demonstrated that two additional methods of contiguous spread exist once tumor involves the middle ear mucosa. The medial wall of the eustachian tube and middle ear are separated from the carotid artery by a thin bony plate that, in some areas, is less than 1 millimeter thick. In temporal bone specimens, this thin bony partition had been penetrated by tumor that had involved extensively the sympathetic nerve plexus surrounding the carotid artery. In addition, tumor had penetrated the posterior mastoid air cells and reached the dura. From the dura, tumor had subsequently spread to the internal auditory canal. Direct extension into the labyrinth can also occur but is considered unusual (Rosenwasser, 1940; Hiraide et al, 1983). High local recurrence rates are not doubt due to the invasive nature of this disease along these multiple pathways.

Histologically, most lesions are reported to be well or moderately well differentiated (Friedman, 1974). No difference has been noted in terms of survival with respect to the differentiation of the tumor in the past (Johns and Headington, 1974; Arthur, 1976). In a small series of cases, Kenyon and colleagues (1985) reported a marked trend toward improved survival in poorly differentiated lesions treated with radiotherapy. For the moment, however, this must be taken as an isolated reported that requires further confirmation.

Clinically, there are not pathognomonic signs or symptoms of squamous cell carcinoma of the external ear canal, middle ear, and temporal bone. The majority of patients will present with a history of prolonged aural discharge, which may have recently become bloody, associated with a conductive or mixed hearing loss. Patients with advanced disease complain of boring pain deep in the ear, which is especially severe at night. Paralysis of cranial nerve VII is characteristically slowly progressive and has been reported to occur in up to one-half of all cases. Over one-half of patients will relate a history of chronic suppurative ear disease of 20 to 30 years' duration. Regional nodes are generally not involved with disease and are positive in only 10 to 15 per cent of cases (Barnes and Peel, 1985; Nadol and Schuknecht, 1984; Wagenfeld et al, 1980). Wang (1975) has offered survival data for planned postoperative radiotherapy combined with complete mastoidectomy, which is comparable to survival data offered for more extensive surgerical resections combined with postoperative radiotherapy. Most likely, in this study, owing to the advanced infiltration of these lesions at the time of clinical diagnosis, there has not been a clear spearation of survival data between those patients treated with partial temporal bone resections and radiotherapy, and those patients treated with complete mastoidectomy combined with postoperative radiotherapy.

Mastoidectomy with open cavity techniques followed by planned postoperative radiotherapy has entailed significant morbidity in the past, because of the relatively high percentage of patients who developed osteoradionecrosis. Nadol and Schuknecht (1984) have demonstrated, however, that with muscle flap obliteration of the mastoid cavity, the incidence of osteoradionecrosis can be markedly reduced.

The authors' treatment for squamous cell cancer limited to the external ear canal is a lateral temporal bone resection. In the treatment of extensive lesions, this resection is combined with planned postoperative radiotherapy. Routinely, the excision includes resection of a wide field core of concha and tragus. The en bloc canal resection is facilitated by performing a mastoidectomy with an extended facial recess approach that is modified to permit removal of the entire external ear canal and tympanic membrane. In selected cases, a modified neck dissection and lateral parotidectomy are performed simultaneously. When the facial nerve is free of tumor, it is not sacrificed. The resulting mastoid and middle ear cavity is primarily grafted with a thin split-thickness skin graft. Fascia temporalis is used to reconstruct the tympanic membrane. Complete healing can be expected with this technique in 4 to 6 weeks, following which radiotherapy, if indicated, is begun. More extensive temporal bone resections are employed in lesions involving the mastoid portion of the middle ear. Cancers involving the anterior middle ear space are difficult to adequately resect because of their proximity to the carotid artery. In such instances, the authors have performed a canal wall down mastoidectomy followed by postoperative radiotherapy as an alternative to an en bloc temoral bone resection.

It is hoped that in the future, multi-modality therapy, along with earlier diagnosis, may offer improved survival statistics for this disease.

Verrucous Carcinoma

Verrucous carcinoma, also known as Ackerman's tumor, is a rare and poorly understood lesion of the temporal bone. Although it has been considered a variant of squamous cell carcinoma, verrucous carcinoma has distinct biological and clinical characteristics. The head and neck are the most common location of this tumor. Most cases occur in the oral cavity and larynx; less common sites include the nose, paranasal sinuses, and very rarely the temporal bone.

Ackerman first described verrucous carcinoma of the oral cavity in 1948. He described its gross appearance as a fungating, warty, papillomatous lesion. Histologically, these tumors have deeply projecting, cleftlike spaces, with advancing margins of abundant inflammatory cells. Microscopically, because of their high degree of cellular differentiation and infrequent cytologic atypia, they are often considered nonmalignant. However, their local destructiveness and propensity for local recurrence classifies them as truly malignant lesions. Ackerman also noted that regional lymph node involvement and distant metastases were rare.

In 1981, Woodson and colleagues documented the first case of a middle ear verrucous carcinoma; in 1984, Proops and associates reported a second case. Five additional cases were reported by Edelstein and co-workers in 1986. Like squamous cell carcinoma of the temporal bone, the peak incidence of verrucous carcinoma of the temporal bone is in late middle age or older.

The original case presented by Woodson was a 72-year-old man with facial paralysis and more than a 20-year history of otorrhea. It was initially thought that the patient had an "unusually aggressive cholesteatoma". An erosive lesion of the mastoid, which extended to the middle cranial fossa causing multiple nerve paralyses, was eventually found. He was treated with many surgical procedures, with subsequent radiotherapy. Proops and colleagues presented a similar patient with chronic mastoiditis and an epithelial, papillomatous lesion of the mastoid. He was treated by mastoidectomy followed by combined chemotherapy and radiotherapy. Edelstein and co-workers' series had similar presentations. Three of their five patients were treated surgically and had favorable outcomes. Two patients had unusually aggressive verrucous cancers that failed combined treatment; one of these patients had surgery and chemotherapy, and the other had surgery and radiotherapy. In this last series of five patients, the survival period ranged from 6 months to over 7 years.

As with any newly described disorder, many questions remain unanswered regarding verrucous carcinoma of the temporal bone. The authors speculate that chronic infection may be the stimulating cause. Malignant degeneration of a pre-existing cholesteatoma may also have occurred in some cases. Woodson believes that this tumor may really be more common and be misdiagnosed as "aggressive cholesteatoma" or as a slow-growing variant of squamous cell carcinoma.

A detailed clinical history must be furnished to the pathologist if an accurate diagnosis is to be made. The use of radiotherapy remains debatable, since some authors speculate that

this may cause anaplastic transformation of verrucous carcinoma.

Primary Adenomatous Lesions

Primary adenomatous lesions are uncommon tumors of the middle ear. Traditionally, these tumors have had many names, including cystadenomas, ceruminous adenocarcinomas, pleomorphic adenomas, and glandular tumors. Recently, the consensus has been to limit the classification to adenoma, adenocarcinoma, and adenoid cystic carcinoma. In 1980 Dehner and Chen added a fourth possible group, pleomorphic adenoma.

Historical Review

In 1904, Lange reported an adenocarcinoma involving the middle ear in an 83-year-old patient with long-standing otorrhea, pain, aural polyps, and facial nerve paralysis. Furstenberg in 1924 reported a primary adenocarcinoma involving the middle ear and mastoid and emphasized that the most prominent symptom of cancer of the middle ear is pain. He also stated, as did Newhart (1917), that invasion of the temporomandibular joint is not an uncommon symptom. Furstenberg performed radical excision of the tumor, followed by roentgen therapy. The tumor was regarded as a primary adenocarcinoma of the middle ear, in the absence of any evidence of a primary lesion elsewhere. Furstenberg believed "that the growth, therefore, represented a teratoid disturbance of development with blastomatous transformation". This patient survived more than 4 years. Additional cases of adenocarcoma involving the middle ear have been reported by Heschelin (1925), Malan (1926), Gritti (1934), Siedentrop and Jeantet (1961), Dehner and Chen (1980), and Pallanch and co-workers (1982).

In the early reported cases of adenocarcinoma there was doubt as to whether the tumors arose from the middle ear or from the external canal. Malan's case arose primarily from the mucosa of the hypotympanum. Gritti's case originated from the mucosa near the eustachian tube. However, because of the occasional finding of a simultaneous tympanic membrane perforation, with granulation occurring on both sides, the doubt remained. Even Dehner and Chen's cases of adenocarcinoma arose from both the middle ear and the external canal. Finally, in 1976, Hyams and Michaels established that adenomatous tumors can arise from the respiratoy epithelium of the middle ear, as opposed to the routine adenoma, which arises from apocrine glands elsewhere in the body.

A second question that developed historically was whether there were different grades of adenocarcinoma to fit the various histologic and invasive characteristics of the tumor. Siedentrop and Jeantet in 1961 reported three cases of adenocarcinoma, none of which had metastases, although one had aggressive local invasion and responded poorly. Dehner and Chen presented five cases of adenocarcinoma, of which three were high-grade, with survival of less than 1 year. In 1976, Derlacki and Barney helped to rectify part of the controversy by reporting on three cases of benign adenomatous lesions of the middle ear. Pathologically, these tumors were grayish-tan and polypoid with bright shiny surfaces. Histologically, they had an infiltrative microscopic pattern and cellular variation that seemed to imply adenocarcinoma but lacked any anaplasia or bony erosive areas. Derlacki and Barney implied that these lesions were a different type of tumor. Also in 1976, Hyams and Michaels at the Armed Forces Institute of Pathology presented a series of 20 additional primary adenomatous tumors of the middle ear.

In 1984, Eden and colleagues presented four cases of adenomatous lesions of the middle ear. They surmised that what makes these lesions different from more aggressive lesions is that there is no vascular invasion, histologic or radiographic bone destruction, or bizarre nuclei or pleiomorphism. The one characteristic of malignancy that they retain is infiltration and invasion of soft tissue.

Clinical Characteristics

Classically, adenocarcinoma of the ear presents as a lesion filling the external auditory canal, associated with severe pain, marked trigeminal nerve symptoms, transitory or complete facial paralysis, and roentgenographic evidence of a destructive process involving the middle ear and jaw. To make a proper diagnosis, a positive biopsy is necessary. Additionally, another primary focus must be ruled out.

In the Mayo Clinic Series of Pallanch and colleagues in 1982, the most prominent clinical symptoms of patients with either adenomas or adenocarcinomas of the middle ear and external ear included decreased hearing; pain in the ear, face, or head; otorrhea; tinnitus; the sensation of fullness; and facial weakness. Less common symptoms were other cranial nerve involvement, vertigo, and hoarseness. One patient had no clinical symptoms, and the tumor was discovered incidentally. Eden and co-workers (1984) reported that the characteristic features of tumors limited to the middle ear included a mass behind an intact tympanic membrane, conductive hearing loss, normal facial nerve function, fullness in the ear, and tinnitus. Lesions confined to the middle ear were less aggressive than those involving the external canal and therefore presented fewer symptoms. They also noted that the average age for patients with adenomatous lesions was 20 to 30 years. Dehner and Chen (1980) reported a slightly older grouping of 40 to 60 years as the period covering all types of glandular tumors of the ear. Since Eden and colleagues' cases were diagnosed at an earlier age, it may explain their patients' improved recovery rate.

One of the interesting facets of this tumor is that it may be present for a long period of time before manifesting symptoms. Similar tumors of the external canal usually become evident earlier because of a less isolated environment. In the series of Pallanch and colleagues, the mean duration of symptoms of progressive conductive hearing loss and recurrent otorrhea and otalgia was 14 years before the correct diagnosis was made.

The differential diagnosis of adenoma-like middle ear masses includes the previously mentioned tumors, as well as metastatic lesions, choristomas, lipomas, meningiomas, glomus tumors, congenital cholesteatomas, and carcinoid tumors. Pallanch and co-workers reported finding one case in the literature of a carcinoid tumor resembling an adenoma of the middle ear. Electron microscopy may help in some cases (ie, differentiating adenoma from adenoid cystic carcinoma).

Treatment

Adenomas of the middle ear are best treated by surgical excision. If the lesion is limited to the middle ear, a simple middle ear exploration may be sufficient. Larger lesions necessitate greater exposure, provided by a mastoidectomy or facial recess approach. Since slow growth and local invasiveness is a hallmark of these lesions, recurrences can be expected

and should be treated in a similar fashion.

Adenocarcinoma can also be treated by wide surgical excision. High-grade forms are often resistant to full excision. Radiotherapy has been tried with mixed success. Stone and associates (1975) reviewed a short series in which he recommended radical mastoidectomy and post-operative radiotherapy. Pallanch and co-workers (1982) noted that this combined treatment often failed if the tumor had invaded the dura or base of the skull or other inaccessible areas.

The adenoid cystic carcinoma has the same histologic appearance and malignant behavior as its salivary counterpart. This lesion is a persistent, aggressive tumor that spreads locally along neural pathways and metastasizes by vascular and lymphatic routes. Therefore, aggressive surgery and radiotherapy is the treatment of choice. Since this tumor has a marked disposition for invasion of neural structures, the facial nerve is usually excised and a cable graft inserted. Nevertheless, this tumor, like its other adenomatous cousins, often has a prolonged clinical course, and long-term survival is possible in many cases. Dehner and Chen (1980) reported that one patient with this tumor was living with pulmonary metastases 28 years after the initial surgical excision. Five-year cure rates are not meaningful in cases of adenoid cystic carcinoma, since recurrence will frequently occur after this time period. Life-long follow-up is essential.

Rhabdomyosarcoma

Rhabdomyosarcoma originating within the temporal bone is a rare neoplasm that occurs almost exclusively in young children. This tumor is the most common soft tissue malignancy in the pediatric population, with 50 per cent occurring in the head and neck. The most common sites, in order, are the orbit, nasopharynx, and ear (Dehner and Chen, 1978). The average age of onset of the tumor is 4 to 5 years, with an equal male-to-female ratio.

The initial presentation of this disease can mimic otitis media. The child generally presents with a recent onset of painless, bloody otorrhea, hearing loss, and a polypoid, friable mass filling the ear canal. Frequently facial paralysis is present, although multiple cranial nerves may be involved. Canalis and Gussen in 1980 differentiated those tumors arising in the middle ear from those with a petrosal origin. The middle ear type undergoes three phases. The first presents with middle ear obstructive symptoms, the second with hearing loss, and the third with "brain, neck and infratemporal and parapharyngeal speace invasion with or without the appearance of metastases" (Deutsch and Felder, 1974). In contrast, the petrosal group demonstrates persistent headaches and abducens and trigeminal nerve paralyses. Biopsy of the lesion will establish the diagnosis. Additional biopsies of the nasopharynx should be taken to evaluate this area as a possible primary source.

Generally, rhabdomyosarcoma of the middle ear is an aggressive, locally invasive neoplasm that tends to metastasize early. Intracranial involvement and extension into the nasopharynx occurs frequently. This involvement gave this disease its traditional reputation for survival rates based in months rather than years. In 1974 Deutsch and Felder reviewed 78 cases reported in the literature and found bone and lung to be the most common sites of metastases. The Intergroup Rhabdomyosarcoma Study (IRS) (Tefft, 1978; Raney et al, 1983) provided a new protocol that prolongs survival to 3.6 years in 47 per cent of 19 patients with

localized disease. Children with meningeal extension still did very poorly. The IRS divides the children into four groups, based on the extent of the disease after surgery.

Traditionally, rhabdomyosarcomas have been treated with a combination of surgery and radiotherapy. In the IRS, patients with full local excision received postoperative vincristine, dactinomycin, and cyclophosphamide. In addition to this treatment, patients with residual disease received radiotherapy. Patients with metastatic disease at initial presentation received all the previous modalities plus adriamycin. Whereas none in the last group survived, the first three groups had varying survival times from 1.5 to 6.5 years. Perhaps Schwartz (1980) put it best when he entitled a paper on this neoplasm " A Wolf in Sheep's Clothing".

Fibrosarcoma

The malignant character of fibrosarcoma is evident in the infiltrating destructive manner of its growth, especially after it has become well established. To decide upon the nature of a connective tissue tumor that, as far as its microscopic morphology is concerned, might be a benign fibroma or a malignant sarcoma, it is essential to know the history of its growth and its gross relations to the adjacent tissues. In one patient the lesion obviously encroached upon the facial nerve and clinically confirmed the pathologic description of fibrosarcoma.

Attacks of transitory paralysis of the facial nerve recurring over a long period should make one suspect a neoplasm. Of further diagnostic significance is a prolonged history of pain in the ear in the absence of aural infection. The presence of a firm tumor deep in the external auditory canal, associated with prolonged pain and evanescent facial paralysis, with or without discharge from the middle ear, should always suggest biopsy to rule out the possibility of malignant disease.

Biopsy in one such patient was made from a hard mass adherent to the posterior canal wall and obscuring the drum. The lesion was reported to be "cellular fibroma with slight cell atypism and sporadic mitosis". Because this examination was not satisfactory, a second biopsy was performed; this revealed "a fibromatous tumor which, however, in one area showed marked cellularity with occasional mitosis, highly suspicious of fibrosarcoma".

Because of the pathologic report and the poor physical condition of the patient, irradiation was decided on as the method of treatment. The local lesion subsided. The patient has been followed for 4 years, and the disease remission represents an unusual result obtained entirely by irradiation.

Hesse said in 1932 that it is not always advisable to undertake dangerous and deforming operations to achieve wide removal of a sarcoma, as one would attempt to do in a case of carcinoma, because sarcomas are more radiosensitive. The result obtained in this instance, although only a single case, is interesting. Keeler (1934) believed that the prognosis for sarcoma involving the middle ear is much worse in young children than in adults.

Osteogenic Sarcoma

Osteogenic sarcoma is a very unusual lesion of the head and neck and is seen only rarely in the temporal bone. Six per cent of these tumors occur in the jaw or maxilla. The main causative factors include trauma, preexisting bone disorders, and irradiation. The pathohistologic features may be osteoblastic, fibroblastic, or chondroblastic. There is no relation between the microscopic appearance and the long-term outcome of sarcoma. Gertner and colleagues (1983) reported one case of osteogenic sarcoma of the temporal bone following previous radiation therapy for a squamous cell carcinoma of the nasopharynx with metastases to the neck. Treatment included temporal bone resection. This tumor was controlled in other areas of the body by chemotherapy and radiation. This is a very rare lesion, and few conclusions can be drawn from this case.

Liposarcomas

Liposarcomas are uncommon sarcomas that occur rarely in the temporal bone. Early diagnosis helps in the treatment and prognosis of the disease. Four types of liposarcoma exist: myxoid, round cell, well-differentiated or adult type, and pleomorphic. The myxoid type rarely metastasizes, whereas, in contrast, the pleomorphic type is less differentiated and is more aggressive. The treatment of choice is wide local excision, although some groups have tried radiotherapy.

Only four cases of liposarcoma in the temporal bone have been reported. The first case, reported by Stout in 1944, was a 37-year-old man with a myxoid tumor in the temporal bone region who had no recurrence 6 years after local excision. Two cases were reported by Enterline in 1960 that had similar results. Agarwal and colleagues in 1975 reported a case in which a 4-year-old boy had a subperiosteal disease of the mastoid masking a liposarcoma.

Radiation-Induced Carcinoma

Malignancies of the middle ear and mastoid secondary to radiation are very rare. Twenty-nine cases have been reported in the literature. Most were due to systemic intoxication by radium, although cases reportedly have been due to external beam therapy. Most radiation-induced tumors of the mastoid are epidermoid carcinomas, although mucoepidermoid carcinomas have been presented as well.

The three groups reported to be at risk for the systemically induced cancers are luminous watch dial workers, patients who were treated with intravenous or oral injections of radium, and industrial workers and researchers involved in the production of radium. Littman and colleagues (1978) reviewed over 5000 people who were "occupationally or therapeutically exposed to radium". They reported 21 cases of mastoid carcinoma, of which 12 were epidermoid, eight mucoepidermoid, and one an unknown variety of cancer. Three of these patients developed sarcomas of the bone outside the head and neck region. Tumor induction may be caused by radon gas trapped in the mastoid air cell system or from direct concentration of radioactivity in the temporal bone.

A larger potential source of radiation-induced carcinoma is the expanding pool of patients who receive radiotherapy to the head and neck. This risk is considered to be

extremely samll. Ruben and associates (1977) presented one case of epidermoid carcinoma of the mastoid following a total of 14.701 rads to the posterior fossa over a 12-year period. The patient had an astrocytoma that had failed to be removed at three craniotomies. Weshler and co-workers (1983) presented a case of a patient who had received 9700 rads to the neck and temporal bone for papillary adenocarcinoma of the thyroid and recurrent neck disease. The dose of radiation in both these cases was excessive.

Patients with radiation-induced carcinoma of the temporal bone presented with chronic inflammatory ear disease that frequently masked the correct diagnosis. Radiologic evaluation revealed destruction of the bony walls of the mastoid, not unlike that seen on x-ray studies of chronic mastoiditis and cholesteatoma. The latency period from the time of initial exposure to the time of diagnosis averaged 30 years, with Littman and colleagues (1978) presenting latencies of 21 to 50 years.

Treatment of radiation induced carcinoma includes temporal bone resection and radiotherapy. Littman and co-workers reported two patients surviving 14 and 21 years following combined therapy. The use of radiotherapy in these patients would seen paradoxical. Perhaps the most important aspect of this rare disease is that a large number of patients are potentially at risk.

Invasion of the Middle Ear, Mastoid, and Temporal Bone by Tumors from Adjacent Areas

Metastatc Tumors

A patient with a known primary cancer who develops ear symptoms should be suspected of having metastatic spread to the temporal bone. Frequently, a patient who has no history of ear disease will present with symptoms resembling acute mastoiditis and during surgery be discovered to have metastatic disease from an occult primary tumor. The most common primary tumors that one should look for are lung, breast, kidney, and prostate.

Five different patterns of metastatic involvement of the temporal bone were described by Berlinger and colleagues in 1980. The most common type is due to isolated metastases from a distant primary tumor such as adenocarcinoma of the breast. The route of invasion is the marrow-containing spaces of the petrous portion of the temporal bone, which may filter out malignant cells from the blood. Another common route for metastasis is direct extension from the nasopharynx. This may occur in squamous cell carcinoma of the nasopharynx, which may erode the petrous apex and invade along the eustachian tube. Other patterns of metastasis may include cerebrospinal fluid spread, direct extension from an intracranial tumor, and leukemic/lymphomatous infiltration.

The basic approach to potential metastatic lesions is a combination of clinical workup, relevant x-ray studies, and local ear biopsies. If a physician strongly suspects a metastatic lesion, then specific studies of abnormal suspected organ systems can be performed prior to any ear manipulation. CT scannnig of the temporal bone may reveal a destructive bone lesion that is more suggestive of metastases than an expanding growth in the temporal bone. Technetium bone scanning of the whole body may reveal suspicious areas other than the ear or skull. Routine blood and urine tests may help focus the physician on a potential primary

medical problem. Standard chest x-rays may be helpful in the patient who smokes and has a chronic cough. Biopsies of aural polyps or granulation tissue may help reveal primary sites. Only when the basic clinical evaluation fails or simple biopsies prove nondiagnostic, should more extensive procedures (eg, mastoidectomy) be performed to gain access to a lesion or to obtain additional tissue for study.

The treatment of metastatic ear lesions depends not only on the severity of the symptomatology but, more important, on the treatability and prognosis of the primary lesion. In the past, otitic lesions were treated with surgery or irradiation. At present, some breast and prostate cancers are treated with combined hormonal therapy and chemotherapy. These approaches may also help control metastatic sites in the ear.

The reported incidence of metastases to the temporal bone is small; however, this may be misleading. In a patient riddled with cancer, metastases to the ear may not be diagnosed unless the patietn develops ear symptoms. Therefore, occult spread to the temporal bone may occur more frequently than has been reported but may be overlooked because of the life-threatening or debilitating effects of the primary cancers. The otic capsule is usually resistant to metastatic lesions, although vertigo and hearing loss may indicate neural carcinomatosis. Metastatic lesions should be included in the differential diagnosis whenever standard therapies of routine ear problems fail. For example, metastases should be considered whenever the ear bleeds profusely following a biopsy or when a patient has a progressive facial weakness with no known cause.

In 1982, Parisier and associates studied a subgroup of patients with metastatic ear lesions who had facial nerve paralysis. The most commmmon primary sources, in order of frequency, were lung, breast, kidney, and colon, with isolated cases arising in skin, larynx, ovaries, testes, adrenals, and cecum. In most cases, the facial paresis was a progressive one that became complete and therefore was clinically distinct, and it should never be confused with Bell's palsy. When the neural involvement was caused by compression, shrinking the lesion with appropriate therapy resulted in improved facial movement. However, if the nerve was invaded by tumor, the paralysis was not reversible.

In recent years, multiple paraneoplastic syndromes have been described regarding the central nervous system and peripheral nerves. McGill presented an interesting case that bears mentioning in this section on metastases, even though no direct infiltrates were found in the ear. The patient was a 54-year-old woman with oat cell carcinoma of the lung, which caused carcinomatous encephalomyelitis and resulted in sudden deafness and vertigo. The pathologic temporal bone findings included almost total loss of cochlear neurons and degeneration of the vestibular nerve. This case presented an otologic neuropathy secondary to a primary neoplasm.

Lymphoma and Leukemia

Lymphoma may involve the temporal bone. It was first reported by Proctor and Lindsay in 1947. In 1972, Paparella and el-Fiky presented the largest series, which included eight patients with malignant lymphoma. The three forms of lymphoma found in the 16 temporal bones from this study included lymphosarcoma, reticulum cell sarcoma, and Hodgkin's disease.

The route of invasion of lymphoma into the ear most commonly is through the narrow spaces or by infiltration into the perilymphatic space around the eustachian tube. This accounts for the high incidence of otitis media, tympanic membrane scarring from chronic infection, and hearing loss in Paparella's cases. Also, perivascular infiltration of the vestibular and facial nerve caused progressive hearing loss and facial paralysis. In one patient, involvement of the scala tympani occurred, which caused sensorineural hearing loss. Treatment should be focused on the general systemic problem in the patient (eg, chemotherapy) with a conservative approach aimed at relieving local ear symptoms (eg, antibiotics or myringotomy).

Meningiomas

Meningiomas represent 13 to 19 per cent of intracranial neoplasms. The majority are supratentorial, and less than 10 per cent occur in the posterior fossa; of these, approximately one-half occur on the posterior surface of the temporal bone. Asymptomatic meningiomas have been found at autopsy to have an incidence of 1.4 per cent. These benign tumors are more common in women by a ratio of 2:1 and have a peak incidence of 45 years of age, according to Nager, who has pursued a lifelong interest in the subject of meningiomas (Nager et al, 1983).

Meningiomas arise from arachnoid cells that cluster around arachnoid villi. These villi accompany the major dural sinuses and are found at the foramina of the skull base and also occur in ectopic clusters within the temporal bone. Nager (1964) and Guzowski and co-workers (1976) have demonstrated ectopic placement of arachnoid granulations along the greater petrosal nerve, in the internal auditory canal, at the geniculate ganglion, and in the jugular foramen within the temporal bone. These ectopic arachnoid granulations are felt to be the origin of primary temporal bone meningiomas.

However, the majority of meningiomas found within the temporal bone are most likely secondary to intracranial meningiomas of the posterior fossa. Since intracranial meningiomas are frequently clinically silent until they are quite large, the otologic symptoms may predominate, obscuring the intracranial pathology. Parisier has emphasized that although careful neurologic evaluation had been normal in three of his patients diagnosed as having primary intratemporal meningiomas, with the advent of CT, all three were found to have relatively large posterior fossa meningiomas (Parisier et al, 1978). He concluded that any patient having a middle ear meningioma should be highly suspect for an intracranial meningioma until this is ruled out by a high-resolution CT scan with intravenous contrast.

Once a meningioma has gained access to the middle ear, it may present as a middle ear mass, an aural polyp, or mass or granulation tissue, coming through a perforation. Secondary infection is common. Conductive hearing loss predominates and, according to Proctor and Lindsay (1947), meningiomas almost never invade the bony labyrinth. Sensorineural hearing loss, however, may predominate if the lesion involves cranial nerve VIII in the posterior fossa or internal auditory canal. Additional clinical symptoms may include headaches, tinnitus, vertigo and cranial nerve VII paralysis or weakness.

Histologically, the tumor is made up of masses of endothelial cells, the nuclei of which are large and uniform in size and in whorls. As stated by Nager (1964), these tumors

contain round psammoma bodies made up of concentric layers of hyalinization and calcification. There may be considerable fibrous tissue in some tumors, and in others there may be many vascular spaces. If only limited biopsy specimens are available, these lesions may be confused with glomus tumors. Both tumors have nests of cells that tend to be uniform. However, an epithelioid appearance of the cells in a highly vascularized stroma suggests a diagnosis of glomus tumor. Reitz and colleagues (1983) demonstrated that, if necessary, these lesions can also be clearly differentiated by electron microscopy. Meningiomas are characterized by this technique in that they present with elaborate cytoplasmic processes that interdigitate in an interlocking jigsaw pattern. Computerized axial tomography has been extremely reliable in detecting intracranial meningiomas. It permits accurate observation of areas of calcification that may be present within the meningioma, as well as visualization of the hyperostotic changes that may be present in the calvarium adjacent to the tumor. Since meningiomas are usually vascular lesions, contrast infusions generally produce enhancement that facilitates the visualization of this tumor.

Primary intratemporal meningiomas are quite rare. Several cases have been documented by Nager (1964), Guzowski and associates (1975), Salama and Stafford (1982), and Reitz and co-workers (1983). Long-term follow-up of any lesion deemed to be of primary temporal bone origin is necessary in order to rule out the later appearance of an intracranial meningioma, which may have been the primary site of origin.

It is the consensus of those who have experience with this neoplasm that surgical removal is the treatment of choice. If the neoplasm arises in the temporal bone, the cure rate will be greater than when the tumor rises from outside the temporal bone and contiguously invades it. Some cases require a combined neurosurgical and otologic approach to effect a cure. Irradiation is not regarded as especially effective in the treatment of meningioma.

Gliomas

Gliomas are central nervous system tumors and, according to Nager (1967), they may be classified into four categories: neuroepithelial tumors, mesodermal tumors, ectodermal tumors, and congenital and embryonic tumors.

The group of neuroepithelial tumors includes medulloblastomas, gliomas, paragliomas, and gangliocytomas. It is worthy of note that Nager reported five cases that involved the temporal bone by direct extension or metastatically. Of interest is Nager's comment that the morphology of the tumor does not change as it spreads and invades the temporal bone.

Cylindroma

Cylindromas is present most frequently in the salivary glands, and occasionally in the lacrimal glands and mucous glands of the upper respiratory tract. As a rule, it is a slow-growing tumor. There is a characteristic tendency for the tumor to recur after surgical removal. Distant metastases to the lungs and to bone are not unusual late in the course of the disease.

Cylindroma of the parotid gland with extension to the middle ear and mastoid portion of the temporal bone, fortunately not common, is a rather hopeless situation. Combined

surgical and radiation therapy and chemotherapy have little to offer in advanced cases. Early recognition, which establishes the diagnosis while the lesion is still localized, offers the best change of cure with surgery. Even in these instances the ultimate prognosis must be guarded.

Dermoid Cyst

Dermoid cyst is a simple form of teratoma that may occur almost anywhere. Generally a dermoid cyst is round or irregularly multilocular and when opened is found to have a tough, hard wall; it is often filled with a soft, greasy mass of granular, buttery consistency in which hair may be found. There may be several sebaceous glands that secrete the thick, greasy substance. The mass frequently contains growth muscle, fat, and connective tissue. On occasion, teeth have been noted in the cyst. One such dermoid cyst was operated upon by Proud and Kirschner; the lesion, located adjacent to the mastoid process, had enlarged and eroded into the mastoid bone.

Tumors of the Nasopharynx

Tumors of the nasopharynx may spread to the middle ear and temporal bone via the eustachian tube, petrous pyramid, or along the base of the temporal bone. According to Bloom (1961), tumors of the nasopharynx are highly malignant and most are undifferentiated. There is a wide spectrum of variation in the degree of anaplasia in different areas of nasopharynx tumors in different patients and at different times in the course of the disease. The tumor may vary in appearance from the primary site to the metastasis. These tumors have a great tendency to spread via lymphatic channels and to involve regional lymph nodes early. In the late stages, liver and bone may be involved. There was radiographic evidence of destruction of the apex of the petrous pyramid in 9 per cent of the patients reported in the Bloom series.

The jugular foramen syndrome is not uncommon, indicating involvement of the undersurface of the temporal bone. This may be from tumor extension or from pressure from involve lymph nodes. We have seen what was regarded as a transitional cell carcinoma in the middle ear before the primary lesion in the nasopharynx was established.

A most interesting and unusual example of spread of tumor from the ethmoid and sphenoid regions invading both temporal bones was reported by Shambaugh and Hagens in 1929. According to the authors, the diagnostic possibilities were (1) basal cell carcinoma and (2) endothelioma. They stated that the region of the eustachian tube on each side offered the path of least resistance and that growth was most rapid along this route into the middle ear.