Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 5: Skull Base

Chapter 55: Surgery of the Cranial Base

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The last 20 years have seen more advances in surgical management of large lesions of the skull base than in any other area of neurotologic surgery. Increasing cooperation between neurosurgeons, otologic surgeons, and head and neck oncologic surgeons has rendered lesions previously considered unresectable suitable for operative removal. The participation of a variety of specialists in internal medicine has permitted satisfactory postoperative recovery in patients previously deemed inoperable.

Pathology and Pathophysiology

Benign Lesions

Neuromas

Schwannomas may affect cranial nerves VII through XII and may present as skull base lesions. Schwannomas of cranial nerves VII and VIII (acoustic neuromas) are discussed in Chapter 54. Neuromas of cranial nerves IX through XII occur uncommonly. Early diagnosis is important because, despite their nonmalignant nature, these lesions can cause death from central nervous system invasion and profound morbidity from dysfunction of cranial nerves IX through XII. Schwannomas of the inferior cranial nerves are generally found as isolated lesions but may be associated with other neuromas or meningiomas in patients with von Recklinghausen's disease. Neuromas of the jugular foramen frequently involve young patients.

The most common mode of presentation is dysfunction of a single lower cranial nerve; therefore, the patient may complain of dysphasia or change in the quality of his or her voice. In patients with neuromas of the hypoglossal nerve, asymptomatic deviation and fasciculation of one-half of the tongue may be the only abnormality. As the tumor grows, the other cranial nerves passing through the jugular foramen will become involved, and a complete jugular foramen syndrome may result. Patients who have involvement of both cranial nerves IX and X will invariably experience significant problems in swallowing. Chronic aspiration is typical of this population, and many experience notable weight loss.

The next most common presenting symptom is unilateral neurosensory hearing loss as a result of compression of the contents of the internal auditory canal (IAC) from below. The ipsilateral auditory brain-stem response (ABR) is therefore abnormal. Absolute wave V latency

and III-V and I-III interpeak latencies will be prolonged.

Headache from posterior fossa invasion, Horner's syndrome from compression of the cervical sympathetic ganglia, disequilibrium, tinnitus, and a palpable mass high in the neck sometimes accompany the presenting symptom.

Meningiomas

Meningiomas, although less common in the posterior fossa, do arise with regularity. Meningiomas, although less common in the posterior fossa, do arise with regularity. Most remain intracranial, but a significant percentage (around 20 per cent) will erode through the skull and present extracranially. Involvement of the temporal, infratemporal, and pterygomaxillary fossae usually requires a skull base approach combined with intracranial surgery in order to assure complete removal. "En plaque" meningiomas will present special problems because of the rather diffuse involvement of large areas of dura. The postoperative care of these patients is frequently complicated by cerebrospinal fluid leaks, because significant amounts of dura usually will need to be resected.

Cholesteatoma

Cholesteatomas arise from the congenital rest of squamous epithelium within the skull. They are most common with the temporal bone. Histologically they are indistinguishable from epidermoid inclusion cysts, and they accumulate keratinous debris and desquamated epithelial cells within their capsules. They grow slowly, with bony erosion secondary to the effects of pressure and lytic enzyme activity. Because of their slow rate of growth, neural and vascular structures are often able to accommodate to significant displacement and distortion. These lesions are therefore often of very large size prior to the onset of symptoms. Congenital cholesteatomas may present with unilateral sensorineural hearing loss, tinnitus, or disequilibrium due to compression of cranial nerve VIII within the IAC or cerebellopontine angle. The ABR is therefore usually abnormal.

Hypesthesia of one or more divisions of the trigeminal nerve is more common with congenital cholesteatomas than with other temporal bone neoplasms or skull base lesions. Paralysis of cranial nerve VI, with resultant diplopia, is rather frequent and is a consequence of involvement at the petrous apex. Headache and hydrocephalus are also common at the time of presentation.

Cholesterol Granuloma

Chemodectomas

Chemodectomas arise from glomus bodies distributed along parasympathetic nerves in the thorax and neck and at the skull base. Glomus bodies are made up of chemoreceptor tissue capable of secreting hormonally active monamine peptides. They derive from cells of the neural

crest. They are believed to play a role in the compensatory physiologic response to hypoxia, hypercarbia, and acidoses. Histologically the glomus body consists of a fibrous stromal septae that divide the gland parenchyma into lobules called glomeruli. They are profusely vascular. Within the temporal bone, glomus bodies may be found within the adventitia of the jugular bulb along any part of cranial nerves IX and X, including the tympanic canaliculus, retrofacial air-cells, cochlear promontory, and geniculate ganglion. In the neck, they are found at the carotid body, within the nodose ganglion (vagal body) along the superior and inferior laryngeal nerves and within the mediastinum,

Glomus tumors are the most common benign neoplasms arising within the temporal bone. They manifest considerable variability in their clinical behavior. They may pursue an indolent, protracted course with marked bony erosion, or bony erosion may be relatively absent, with expansion only along naturally occurring tissue planes. A minority of tumors exhibit aggressive growth characteristics, with rapid erosion of bone and short-term survival. Between 5 and 20 per cent of patients present with multiple, syntachronous tumors, and all patients with glomus tumors should be evaluated for multiple lesions. All glomus tumors have the potential for secreting vasoactive catecholamines, although few manifest clinically significant hypersecretion. Norepinephrine-, epinephrine-, and dopamine-secreting tumors have been described.

Symptoms, when present, are related to catecholamine intoxication and include headaches, sweating, palpitations, hypertension, and pronounced anxiety. Symptoms may be relatively constant or occur in paroxysms. The presenting symptoms will depend on the point of anatomic origin and the size of the lesion. Tumors confined to the middle ear space will present with pulsatile tinnitus and conductive hearing loss. Facial paresis or paralysis may occur as the tumor enlarges and be a consequence either of bony erosion or a natural dehiscence from the fallopian canal. Erosion into the membranous labyrinth produces vertigo and neurosensory hearing loss. Drainage from or polyps of the external auditory canal occur when tympanic membrane involvement develops and usually heralds secondary infection.

Tumors that originate in the hypotympanum or from the jugular bulb are more likely to present with signs and symptoms secondary to paresis or paralysis of cranial nerves IX, X, XI, or XII. However, many chemodectomas originating from the jugular bulb area present with middle ear findings only. Tumor involvement of the hypotympanum and jugular bulb area may be extensive even in the absence of cranial nerve involvement. Therefore, even patients presenting with signs and symptoms limited to involvement of middle ear structures need to be fully evaluated for extension of tumor into the inferior portion of the temporal bone.

Malignant Tumors

Primary malignant lesions of the skull base are uncommon. Occasionally, malignant variants of the benign tumors discussed above do occur, and their diagnosis will depend on histologic examination. The management of malignant glomus tumors, malignant meningiomas, and malignant schwannomas is controversial and difficult. Each case should be evaluated individually. Hemangiopericytomas occurs sporadically and have relatively high recurrence rates

even when histologically benign. According to Batsakis, the diagnosis has never been made preoperatively. Metastatic lesions of the temporal bone develop with such infrequency that operative removal will be indicated only rarely. Intracranial malignancies and primary carcinomas of the neck may reach the temporal bone by direct extension. Cutaneous malignancies, primarily basal cell and squamous cell carcinomas that arise from the skin of the external auditory canal and ear, may involve the temporal bone and skull base. Leukemic and lymphomatosis infiltrates occur, and operative intervention may be warranted, especially if facial paralysis is present.

Vascular Lesions

A variety of vascular lesions occur and present great hazards to the unwary. Inadvertent biopsy of these lesions may be fatal. Aberrations of the internal carotid artery require arteriography for definitive diagnosis but may be suspected from computed tomography (CT) scanning. A high jugular bulb may appear as a neoplasm behind the inferior portion of the tympanic membrane. Aneurysms of the internal carotid artery may arise either as congenital defects in the walls of the carotid artery or as a consequence of temporal bone trauma. A-V malformations are not uncommon, and occasionally operative extirpation will be mandated because of intracranial involvement, infection, high output failure, or gross cosmetic deformity. We do not feel that unilateral hearing loss of any magnitude warrants the risk inherent in surgical management of large A-V malformations.

Diagnosis

Cranial Nerve Dysfunction

Paralysis, paresis, or manifestations of irritability of the cranial nerves are the most frequent symptoms of skull base lesions. Analysis of which cranial nerves are involved, coupled with a good understanding of the three-dimensional anatomy of the skull base, will often permit fairly precise localization of the tumor prior to diagnostic imaging.

Involvement of cranial nerves III, IV, or VI results in diplopia. These nerves are susceptible when the lesion involves the base of the middle cranial fossa, cavernous sinus, or petrous apex. Cranial nerve VI is most commonly involved, because its long course across the petrous apex and cavernous sinus makes it the most vulnerable. Meningiomas and aneurysms of the anterior vascular system are the most common causes, but large cholesteatomas of the petrous apex, as well as chemodectomas extending into the cavernous sinus, may produce diplopia.

Cranial nerve VI dysfunction occurs frequently as a result of trauma and basilar skull fracture. It is part of classic triad of Gradenigo's syndrome (cranial nerve VI palsy, retro-orbital pain, and purulent aural discharge) that results from chronic infection of the petrous apex.

The large size and central position of the gasserian ganglion and the long course of the three peripheral divisions of the trigeminal nerve make it vulnerable to a variety of skull base lesions. Meningiomas of the basilar dura of the middle fossa and any large lesion involving the

cerebellopontine angle may produce dysfunction of the trigeminal nerve. Manifestations of irritability in the form of facial pain syndromes are common. Classic tic douloureux is usually caused by vascular compression, but other types of chronic facial pain occur with involvement by meningiomas and/or primary cholesteatomas. Large acoustic neuromas are more likely to result in anesthesia or hypesthesia of the face. If more than one division of the trigeminal nerve is involved or if other cranial nerves are dysfunctional, then it may be possible to locate the area of tumor origin or involvement.

The facial nerve has a long course through the skull base before ramifying within the parotid gland. Disorders of the facial nerve may be due to pathologic processes arising within the brain itself, within the cerebellopontine angle, within the temporal bone, or within the parotid gland. Within the brain, the facial nerve may be affected by vascular lesions, primary brain stem neoplasms, or demyelinating disease.

Despite its proximity to cranial nerves V and VIII within the cerebellopontine angle, the facial nerve demonstrates altered function as a result of mass lesions within the cerebellopontine angle less commonly than do cranial nerves III and V. Therefore, it is said to be less susceptible to the effects of extrinsic pressure within the cerebellopontine angle. However, large lesions within the cerebellopontine angle do occasionally affect the function of the nerve, usually as a late finding. Large acoustic neuromas (> 4 cm) will occasionally produce facial weakness, as will meningiomas. Primary cholesteatomas are more likely to produce manifestations of irritability in the form of facial twitch or hemifacial spasm. Irritability may also be a consequence of vascular compression at the root entry zone analogous to tic douloureux.

Tumors involving the temporal bone - principally chemodectomas, neuromas, and meningiomas, as well as cholesteatomas - may produce facial paralysis secondary to extrinsic compression or, rarely, invasion. Primary schwannomas of the facial nerve may arise anywhere along its course but occur most commonly within the temporal bone. The patient may complain of facial spasm or twitching. The slow, progressive development of facial paralysis may occur and strongly suggests that tumor involvement is the cause. Not infrequently, a period of nerve irritability is followed by a slow, progressive paralysis. If diagnostic imaging in these circumstance fails to demonstrate a lesion, then surgical exploration should be undertaken.

Dysfunction of cranial nerve VIII is the most common reason for patients with skull base lesions to seek out a physician. Hearing loss may be due to a mass effect within the middle ear cleft, ossicular disruption, invasion of the membranous labyrinth, or compression of the IAC. Routine audiometry can differentiate conductive hearing loss from neurosensory lesions and place the pathologic process in the lateral temporal bone.

If other evidence suggests the presence of a mass lesion and if cholesteatoma can be ruled out, middle ear involvement suggests the presence of a chemodectoma or, less commonly, a neuroma arising from the VIIth nerve or a primary middle ear adenoma.

ABR testing will differentiate involvement of the membranous labyrinth from retrocochlear causes if greater than 60 dB of residual hearing is present. Hearing loss from labyrinthine invasion is suggestive of chemodectoma or basilar meningioma. Less commonly, acoustic neuromas, neuromas of nerves IX through XII, or metastatic lesions will be responsible.

The most common cause of retrocochlear hearing loss is the presence of an acoustic neuroma; however, chemodectomas, other skull base neuromas, and cholesteatomas of the petrous apex regularly produce retrocochlear loss as they enlarge. Occasionally, if the tumor can be removed without damaging the cochlear labyrinth, hearing will improve and the ABR will normalize postoperatively.

Disturbance in balance occurs regularly and may also be caused by direct labyrinthine invasion or by compression of the IAC. Unless it is caused by an acute perforation of the labyrinthine end-organ, the symptoms will be mild because compensation can keep pace with slowly developing dysfunction. This is similar to what is seen in acoustic neuroma. Often patients will be so little disturbed by their labyrinthine dysfunction that they will fail to report it unless specifically questioned. It is not uncommon to discover a completely nonresponsive vestibular end-organ in a patient who has reported no clinical disturbance of balance.

Any or all of the nerves of the jugular foramen may be involved. Indeed, paralysis of the lower cranial nerves is the most common presenting symptom of skull base neuromas and occurs with regularity in large glomus jugulare tumors. Isolated involvement of the glossopharyngeal nerve may be clinically silent, and physical findings are sparse. Deviation of the soft palate toward the involved side may be the only finding. If a brisk carotid sinus reflex can be elicited from the contralateral side, its absence on the involved side is confirmation that the branch of glossopharyngeal nerve innervating the carotid sinus (the sinus nerve of hearing) is involved. Taste will be diminished on the ipsilateral posterior tongue, but this is rarely apparent to the patient.

Paresis of the vagus nerve is usually symptomatic, especially with skull base lesions. Whereas slow onset paresis of the vagus nerve in the neck may permit vocal cord compensation by hyperabduction of the contralateral vocal cord, lesions that interrupt vagal nerve function above the take-off of the pharyngeal plexus will be associated with significant dysphasia. The combination of unilateral vocal cord paralysis with failure of adequate cricopharyngeal opening is never well compensated for and will almost invariably result in significant dysphasia, resulting chronic aspiration, which may cause significant weight loss. On physical examination, laryngoscopy reveals paralysis of the ipsilateral cord in a paramedian position.

Involvement of cranial nerve XI produces paresis or paralysis of the ipsilateral sternocleidomastoid muscle and trapezius muscle. If involvement is long-standing, atrophy of these muscles will be present.

Hypoglossal paralysis produces deviation of the tongue toward the involved side, with atrophy and fasciculations. In and of itself, unilateral tongue paralysis is asymptomatic and

patients may be entirely unaware of it. It may, however, significantly increase chewing, swallowing, and articulation difficulties if combined with paralysis of cranial nerves VII, IX, or X.

Cerebellar dysfunction is uncommon and when present will be due to extensive compression. Frank cerebellar invasion is almost never seen. Trunkal ataxia, central pattern nystagmus, and dysdiadochokinesia may be found.

Pain is a frequent presenting complaint and arises from a variety of causes. Compression of one or more roots of the trigeminal nerve may produce pain in the face or retro-orbital area. Classic tic douloureux occurs rarely, but dull aching pain in the mid-face is frequent. The latter is often mistakenly thought to be caused by sinus disease. Retro-orbital pain, as seen in Gradenigo's syndrome, is the most common form of presentation.

Diffuse, dull, occipital headache suggests posterior fossa compression and hydrocephalus. Retro-orbital, frontal, and temporal headaches are more characteristic of middle fossa involvement.

A disturbing sensation of aural "fullness" or "pressure" will often be reported by the patient as ear pain or headache, and careful history taking may be required to determine its precise nature. The discomfort produced may be quite pronounced, and the patient may be notably distressed by it. The symptom may be caused by a middle ear mass, serous otitis media, endolymphatic hydrops induced by the tumor, or neurosensory hearing loss.

Radiographic Assessment

Thorough, detailed radiographic assessment often permits a preoperative diagnosis and is absolutely essential for operative management. Initial evaluation should consist of plain films, which may be taken in the office. The submentovertex view is the most useful view for evaluation of skull base lesions. It may demonstrate enlargement of the foramina of the skull base or of the IAC. Soft tissue masses can be identified and erosion of some or even all of the skull base may be noted.

Definitive radiographic assessment requires computed tomography (CT) with imaging and, frequently, arteriography. Magnetic resonance imaging (MRI) defines the soft tissue limits of the tumor. MRI is very sensitive to dural invasion and parenchymal involvement. When paramagnetic contrast material is used, thrombi of major (arterial or venous) may be detected.

CT adds the details of the bony anatomy to the image generated by MRI. Detailed (1.0-mm cuts) views of the involved areas should be obtained. Contrast material is used if necessary, but often the MRI scan has provided adequate detail of the soft tissue. Consultation with the radiologist is obtained prior to scanning to ensure that the study visualizes the inferior temporal bone, foramen magnum, and upper neck, with special attention to the details of the labyrinth, jugular foramen, and jugular bulb. Unless diligently searched out, involvement of the jugular bulb

and small areas of posterior fossa invasion are easily missed.

While the role of arteriography has been diminishing, it is still a very important study in assessing skull base lesions. The diagnosis of chemodectoma, angiofibroma, hemangiomas, and other less common vascular lesions, and anomalies may be made preoperatively on the basis of characteristic angiographic appearance. If selective embolization or early surgical control of blood supply is needed, arteriography will demonstrate feeding vessels. Assessment of the intrapetrous carotid artery may significantly affect surgical planning.

When a large skull base procedure is planned and carotid artery involvement is suspected, four-vessel arteriography with cross-compression studies to assess competence of the circle of Willis are essential.

If review of the patient's MRI, CT, and arteriograph suggests that the internal carotid artery may need to be sacrificed, than balloon occlusion testing should be performed. The arteriographer inflates a balloon within the involved carotid artery to completely occlude it. The patient, who is awake, is then serially examined to detect any developing deficit. Visual changes, aphasia, loss of motor function, and sensory changes have been reported. Occlusion is maintained for 20 to 40 minutes unless deficits develop, in which case the test is immediately terminated. Some groups recommend that patients who show no dysfunction be stressed by the induction of hypotension or that measurements of cerebral blood flow be obtained during occlusion, but the usefulness of these additions has not been determined. It is assumed that patients who demonstrate poor results on balloon occlusion testing by developing neurologic deficits will suffer a stroke if the carotid artery is sacrificed. However, one cannot assume that patients who have good results on balloon occlusion testing will tolerate sacrifice of the carotid artery, although their changes are improved.

A significant number of skull base lesions are well known to arise multicentrically. Four-vessel arteriograms may detect contralateral lesions whose presence would alter surgical management. Vascular anomalies presenting as temporal bone lesions will not be detected without angiographic assessment, and inadvertent surgical exploration of such lesions can often be catastrophic.

If the sigmoid sinus, jugular bulb, and upper portion of the jugular vein are not well demonstrated on the venous phase of the arteriogram, then selective, retrograde venography should be performed. The presence or absence of involvement of the sigmoid jugular system will profoundly alter the magnitude of any proposed resection. Selective jugular vein venography will definitively identify the lower limit of tumor extent within the jugular vein. We have seen relatively small chemodectomas extend to the base of the neck within the jugular vein.

By utilizing all the information obtained from multiple radiographic studies, a very precise concept of the tumor's exact size and location can be achieved. A determination can then be made as to whether the tumor is resectable and, if resectable, the operative approach that will be needed. The patient can then be informed as to the operative risk, possible complications,

anticipated recovery period, and probable permanent postoperative deficits.

Management

Four management modalities are available: observation, radiotherapy, embolization, and surgery.

Observation. Observation may be appropriate in elderly patients if it is anticipated that continued tumor growth will not significantly reduce life expectancy or diminish quality of life. Tumor size, location, and growth rate are important variables in making such a determination. Since the potential to metastasize generally affects the decision whether or not to withhold active treatment, a firm histologic diagnosis based on a biopsy specimen is essential if observation is to be selected as the mode of management.

Radiotherapy. The role of therapeutic radiation in the management of skull base lesions depends, of course, on tumor histology and is controversial. It is most useful for malignant lesions in which it may be the primary mode of therapy. The vast majority of skull base lesions, however, are benign. Meningiomas and schwannomas classically are radio-resistant and the preferred treatment modality is surgical. However, consideration should be given to treatment by ionizing radiation for inoperable patients and for those with unresectable tumors.

A few institutions have reported good results with radiotherapeutic management of chemodectomas, despite histologic evidence that the tumor is radio-resistant and that radiation is not curative. We have operated on many patients treated with full-course radiation therapy who developed symptomatic recurrences, due to increasing tumor size. We believe that surgical resection offers the patient the best chance of cure. We have seen many patients in whom the diagnosis of chemodectoma was made on the basis of characteristic angiographic appearance alone and for whom primary radiotherapeutic treatment was urged. Upon surgical exploration a number of the lesions were found not to be chemodectomas. Therefore, we believe that a histologic diagnosis is also a prerequisite for the institution of radiotherapeutic management.

Embolization. Embolization has been used predominantly as a means of reducing intraoperative blood loss. For the most part, its use as a single mode of therapy and its use in combination with radiation therapy have been abandoned. Preoperative embolization by selective catheterization of feeding vessels dramatically reduced operative blood loss. Gelfoam has been the most frequently used material, but many others have been tried. Enthusiasm for preoperative embolization seems to be waning, because significant complications have been reported. Death and stroke may occur from inadvertent contamination of the internal carotid artery with the embolized material. This can occur either as a result of shunting of embolized material through the abnormal vasculature of the tumor or as a result of catheter displacement. Prior to embolization, careful consideration should be given to these possible consequences. Careful study of the arteriogram to determine the amount of shunting through the tumor bed and to assess the anatomy of the external carotid artery and the size and configuration of the branches feeding the tumor will help eliminate patients at higher risk for these complications.

Surgery. Despite the magnitude of the usual resection and despite the significant perioperative morbidity, we believe that surgical resection of skull base lesions offers most patients the greatest chance of cure and the highest quality of life.

Surgical Resection

Perioperative Considerations

These large, prolonged surgical procedures must be well planned. In most circumstances 8 to 16 hours or more of anesthesia will be required. It is important that the attending anesthesiologist be permitted to assess the patient's readiness for prolonged anesthesia well before the expected procedure. The patient should be in optimal condition at the time of resection. Those few patients who have tumor-secreting vasoactive substances will require the careful induction of alpha- and beta-adrenergic blocking agents, starting 2 weeks prior to surgery. Pharmacologic therapy should be based on blood levels of circulating catecholamines and should be instituted in patients with circulating plasma levels greater than twice normal, even if the patient manifests no overt signs of catecholamine intoxication.

Those patients who present with paralysis of the lower cranial nerves and chronic aspiration and dysphasia have special problems. Teflon vocal cord injection should be performed about 2 weeks preoperatively, and vigorous pulmonary toilet with antibiotic therapy, if necessary, should be initiated. As pulmonary therapy proceeds, nutrition should be maximized. Nothing short of unequivocal anabolism at the time of surgery is acceptable, even if this requires tube feeding or intravenous hyperalimentation. Despite the improved pulmonary status such treatment procedures, tracheostomy at the time of the definitive resection is essential even if not otherwise indicated when high vagal paralysis is present.

Vigorous pulmonary toilet should be started immediately after surgery and throughout the postoperative period. This is facilitated by the tracheostomy. Those patients who had functioning vocal cords preoperatively but who lose vocal cord function as a consequence of the surgical procedure should have vocal cord injection performed at the end of the procedure or the following day. Initial injection may be with Teflon if the nerve has been resected, or with Gelfoam if the nerve is anatomically intact.

Surgical nutrition plays a critical role in skull base surgery, and problems in maintaining adequate nutrition may dominate postoperative recovery. Preoperative nutrition may be impaired by chronic aspiration, as discussed above. High vagal paralysis will not permit adequate synchronous cricopharyngeal relaxation and is manifested as cervical dysphasia. Patients with facial and hypoglossal nerve paralysis may have difficult manipulating the food bolus and in initiating deglutition.

Cervical dysphasia as a result of vagal trauma will significantly impair swallowing and produce significant aspiration. It may require weeks of postoperative therapy before oral intake is adequate to sustain anabolism. Poor gastric emptying and prolonged adynamic ileus are well

recognized as a consequence of the high vagal lesions that are frequently produced during skull base surgery. Persistent postoperative vomiting and bloating may persist for weeks postoperatively. The use of a nasogastric or gastrostomy feeding tube in these patients is not helpful, and nutritional support is achieved via a jejunostomy tube. Such a tube is placed most efficaciously at the time of the extirpative procedure.

Anorexia secondary to postoperative depression is present in virtually all patients following skull base surgery, and this will profoundly increase the difficulty in relearning swallowing techniques.

Problems in postoperative nutrition should be anticipated. In the preoperative period, aspiration should be controlled and forced feeding via nasogastric tube begun if necessary. The patient should be given a careful explanation of the anticipated postoperative swallowing difficulties and the importance of maintaining positive nitrogen balance. Visits by the therapist responsible for postoperative swallowing rehabilitation should begin in the preoperative period, so that the patient is thoroughly familiar with the techniques of swallowing that he will need to use postoperatively. Intraoperatively, the placement of a tracheostomy tube and an appropriate feeding tube should be part of the overall procedure. In those patients in whom the vagus nerve is sacrificed, performance of a cricopharyngeal myotomy will significantly improve postoperative deglutition.

Nutrition in the early postoperative period should be instituted by either feeding tube or intravenous hyperalimentation as soon as possible. The slow introduction of oral feeding is begun at about 7 to 10 days only after the patient has recovered completely from the prolonged anesthesia and postoperative gastric retention. Oral feeding is begun very slowly, and each feeding takes place with the therapist in attendance. Only after the patient has been able to consume 2000 to 3000 calories per day for 2 to 3 days is tube feeding stopped.

In addition to therapy directed toward rehabilitation of swallowing, a number of other therapies may be required, including speech therapy. Speech deficits may arise from paralysis of cranial nerves VII, X, or XII, or any combination of these, and may therefore be complex. Patients with postoperative paresis of the facial nerve are routinely given facial nerve exercises. Although the physiologic basis for such exercises may be questionable, patient response is enthusiastic.

Physical therapy to resume ambulation may be needed, not only to overcome the effects of prolonged anesthesia and bed rest but also to compensate for balance disturbance secondary to damage to cranial nerve VIII or to the labyrinth. The early introduction of appropriate rehabilitative exercises will reduce the possibility of long-term, chronic shoulder pain in patients with sacrifice of cranial nerve XI.

Early involvement of an ophthalmologist will greatly facilitate ocular problems. The typical problems with exposure keratitis seen in patients with cranial nerve VII paralysis may be seriously increased if a nerve V deficit is also present. Some patients, especially those with

meningiomas of the floor of the middle fossa, may have extraocular motor deficits as a consequence of injury to cranial nerves III, IV, or VI. Patients with facial nerve deficits should be managed aggressively, using artificial tears hourly during the day, ointment and a plastic eye "bubble" at night, and early, liberal lateral tarsorrhaphy.

Many patients will have had a significant amount of dura sacrificed. These patients therefore will be at risk for cerebrospinal fluid (CSF) leaks. Such a leak may present as CSF rhinorrhea, but it is more likely to manifest as accumulation of fluid behind the skin flap. Accumulation of CSF creates a fluid barrier between the medial portion of the flap and underlying soft tissues, which will prevent healing and revascularization; flap viability is therefore threatened. The earliest manifestation of impaired flap viability will be breakdown of the suture line and persistent escape of CSF to the outside. A subarachnoid, cutaneous fistula may result in uncontrollable contamination of the subarachnoid space with cutaneous and environmental organisms. Overwhelming and intractable meningitis may result. The ultimate outcome for those patients who develop meningitis depends largely on the rapidity with which the process is recognized and therapy is begun. Therefore, any postoperative elevation in temperature and any change in level of consciousness demands immediate evaluation of CSF obtained by lumbar puncture.

CSF leaks may be managed in a number of wars. CSF rhinorrhea will usually require reoperation in order to obliterate the eustachian tube or seal the leak more proximally. CSF leaks that cause large accumulations of fluid under the flap should be treated initially with repeated aspiration, large pressure dressings, and either repeated lumbar punctures or continuous spinal drainage. If these methods fail to control the leak, surgical reexploration and operative reclosure will be required. Once a subarachnoid, cutaneous fistula has developed, removal of CSF from the subarachnoid space is ill-advised, because it carries the risk of "pulling" contaminated CSF retrograde into the subarachnoid space.

Patients who have undergone large skull base obliterative procedures will always require a great deal of emotional and psychological support. The patient's appearance may have been altered drastically by loss of hair, a large cervical defect, and loss of function of cranial nerve VII. The presence of tubes and drains are also very disturbing. There may be loss, or severe impairment, of the patient's most basic bodily functions, including speech, hearing, vision, and the ability to swallow. Especially in the postoperative period, the patient will be dependent on outside resources to meet his or her most basic needs. All these factors combine to alter drastically the patient's self-image and may produce a significant depression. Severe depression should, therefore, be anticipated.

Management begins preoperatively by warning the patient that a postoperative depressive reaction is normal and expected. Postoperatively, the patient's depression should be openly acknowledged and discussed. Many patients will require psychiatric consultation and the judicious use of appropriate antidepressant drugs.

Surgical Procedures

A variety of surgical approaches to the skull base are available, and most often more than one approach is used. Each approach provides access to different areas of the skull base. Therefore, accurate tumor "mapping" is a prerequisite for meaningful preoperative planning. A reliable preoperative surgical plan is essential in informing the patient about the nature of the procedure and its possible sequelae, risks, and complications.

A variety of incisions are available through which one may access the skull base. The Conlay-Lewis incision, developed for total en bloc temporal bone resection, has a reliable vascular supply and a good cosmetic result. It is useful for lesions limited to the temporal bone but provides very limited access to the posterior fossa and infralabyrinthine area. A modified parotidectomy incision that is extended superiorly toward the vertex of the skull is very useful for lesions of the temporal fossa, pterygomaxillary space, and nasopharynx. This incision avoids the external auditory canal completely and thus assures minimal changes in hearing postoperatively; however, it provides only limited access to the temporal bone. Lesions arising from or extensively involving the temporal bone are usually best handled by the "Big C" incision. The incision can be varied to provide greater or less exposure of the posterior skull by varying its most posterior limit, and anterior exposure can be increased by extending the two ends of the incision. The incision can be designed to expose as large a portion of the neck as is needed.

Management of the external auditory canal is a separate issue. In some cases it will be possible to leave the bony posterior canal wall and tympanic membrane intact, but it will be necessary to mobilize the auricle and displace it anteriorly in order to achieve exposure. In such cases, a standard vascular strip incision should be made in the external auditory canal in order to prevent postoperative canal stenosis. In other cases, especially those where the tumor involves the intrapetrous carotid artery, the bony canal wall and tympanic membrane will be removed in order to provide adequate exposure of the anterior portion of the base of the temporal bone. In such circumstances, the external auditory canal should be carefully closed from the underside of the flap. It is important to invert such a closure from the inside out. Closure of the canal should be given consideration as the flap is elevated and sufficient canal skin is left on the flap to assure tight closure. Often conchal cartilage will need to be resected in order to obtain sufficient soft tissue to assure water-tight closure. Failure to close the external auditory canal tightly may result in serious postoperative difficulties. If the dura has been resected, CSF may escape through the external auditory canal. An incompetently closed external auditory canal presents a serious source of bacterial contamination, which may result in infection of underlying tissue, especially fat and muscle grafts placed in the obliterated dead space.

Approaches to the Lateral Skull Base

Facial Recess Approach

The least extensive dissection approaching the lateral portion of the temporal bone is the extended facial recess approach. It provides access to the middle ear space, hypotympanum, and mastoid. It provides adequate access for small tumors limited to the middle ear and mastoid. The area of the jugular bulb, infralabyrinthine area, and retrofacial areas are not exposed. The procedure is begun as a standard tympanomastoidectomy by creating a vascular strip prior to making the postauricular incision. Through a standard postauricular incision, the mastoid is decorticated and all of the mastoid air-cells are exenterated, just as in a simple mastoidectomy. The facial recess is then entered and widely opened. If the tumor extends into the hypotympanum, below the floor of the external auditory canal, dissection may be continued inferiorly by sacrificing the chorda tympani and extending the facial recess inferiorly. Dissection is limited medially by the facial nerve, which is skeletonized. Laterally, dissection is limited by the annulus tympanicus and bony external auditory canal, which should not be violated. If the inferior margin of the tumor cannot be completely visualized through this limited opening, an alternate surgical approach affording wider exposure should be selected. "Blind" removal of tumor from areas of hypotympanum is not only extremely hazardous but is also unlikely to result in total extirpation. Residual tumor left in the hypotympanum may grow for years and become massive in size before clinical recurrence is apparent. Wide exposure is absolutely essential when operating on the skull base. Limited exposure almost invariably results in unnecessary damage to important (or vital) neurovascular structures and/or incomplete tumor removal.

Retrofacial Approach

If tumor involves the inferior portions of the posterolateral temporal bone, if it does not extend anteriorly or beneath the labyrinth, and if it does not involve the middle ear, then it may be adequately exposed and removed by a retrofacial approach. Middle ear and hypotympanic extension may be managed by combining this approach with the extended facial recess approach. Combining these two procedures will leave the facial nerve skeletonized as in a thin bony canal arching across the surgical field. Although the jugular bulb can be visualized by this approach, exposure is not adequate if tumor involves the jugular bulb. Anterior exposure is absent.

The procedure is begun as a simple mastoidectomy. After the sigmoid sinus has been exposed laterally and superiorly, it is followed inferiorly into the air-cell system medial to the facial nerve. One should identify the facial nerve throughout its descending segment with surety prior to dissecting retrofacial air-cells, in order to avert injury to it while assuring maximum exposure. The limits of exposure will be the posterior semicircular canal, the dura medially, and the jugular bulb anteriorly. If there is involvement of the jugular bulb and the involvement is limited to the posterior portion of the bulb, then combining the extended facial recess approach with the retrofacial approach and mobilizing the facial nerve may result in adequate exposure. The facial nerve should be mobilized from the geniculate ganglion to the stylomastoid foramen and displaced anteriorly. Exposure is still very limited, but it may be adequate if infralabyrinthine

and jugular bulb involvement is very minimal. If any doubt exists about whether exposure is adequate, a full infratemporal fossa dissection should be performed.

Infratemporal Fossa Approach

The widest possible exposure of the skull base is afforded by the infratemporal fossa approach. Complete removal of large tumors, with simultaneous control of important neurovascular structures, is possible. The temporal and pterygomaxillary fossae are accessible as fat anteriorly as the lateral rim of the orbit and including the infraorbital fissure and both pterygoid plates. The entire intrapetrous carotid artery can be dissected out from surrounding bone as high anterior medially as the cavernous sinus, which forms a natural limit to dissection. Posterior, the entire sigmoid sinus and jugular bulb may be approached. Medial dissection may be carried out beneath the cochlea and labyrinth out to the petrous apex. The basal portion of the petrous bone may be removed, exposing the dura of the posterior fossa anterior to the sigmoid sinus. Inferiorly, the bone of the foramen magnum occipital condyle and lateral portion of the body of the first cervical vertebra can be removed to expose the dura of the lower medulla and upper cervical spinal cord.

When combined with the surgical approaches to the cerebellopontine angle and middle cranial fossa, tumors with intracranial extension may be managed as a single-stage resection. The procedure requires use of the large C-shaped incision carried well into the neck. The flap is elevated in a plane superficial to the temporalis muscle, which is left intact for later use as a rotation flap. The plane between the sternocleidomastoid muscle and the strap muscles is opened and the carotid sheath is exposed. Cranial nerves X, XI, and XII are identified and tagged. The internal and external carotid arteries and the internal jugular vein are separately identified, and vascular loops are placed around them. The facial nerve is identified as it exits the stylomastoid foramen beneath the digastric muscle and is followed anteriorly. The superficial portion of the parotid gland is removed, and the peripheral ramifications of the facial nerve are dissected out. The larger, more proximal branches are freed circumferentially in order to facilitate mobilization of the infratemporal facial nerve.

A simple mastoidectomy is performed with an extended facial recess. The facial nerve is dissected from the geniculate ganglion to the stylomastoid foramen, and all of the bone is removed from it. The sigmoid sinus is identified and followed anterior-inferiorly to the jugular bulb.

In most cases, involvement of the jugular bulb and/or carotid artery or extension beneath the cochlea will require removal of the bony external auditory canal, tympanic membrane, and ossicles. Significant involvement of the carotid artery, and any tumor extension anterior to the carotid artery, will require either anterior subluxation of the mandibular condyle, condylectomy, or partial mandibulectomy. Complete removal of the condyle and ascending ramus will provide wide access to the pterygomaxillary fossa. Removal of the zygomatic arch or root of the zygomatic arch exposes the temporal fossa broadly. When bone removal has been completed, tumor removal is begun. The facial nerve is carefully freed from the investing soft tissues at the

stylomastoid foramen and transposed into the anterior epitympanum. Tumor is removed from around the carotid artery by developing a dissection plane in the carotid adventitia

Cranial nerves IX through XII are followed cephalad from their points of identification in the neck. If a plane of dissection cannot be identified between the cranial nerves and the tumor and is, therefore, inextricably bound to the tumor mass itself, it may be sacrificed. No effort, however, should be spared to salvage these important structures. If the tumor is tightly adherent to the sigmoid sinus or if it extends intraluminally within the sinus or jugular vein, then the sigmoid sinus, jugular bulb, and upper jugular vein are removed. The proximal sigmoid sinus may be controlled either by packing with Surgical or by ligation. The sigmoid-jugular system is then removed with the attached tumor, since other points of tumor fixation have been freed previously. Immediately following extirpation of the sigmoid-jugular system, packing should be placed into the inferior petrosal sinus.

Intracranial extension, if present, may be removed at this time. Intracranial extension frequently involves the posterior fossa. Posterior fossa dura anterior to the sigmoid sinus will be the usual point of penetration, and this area is easily exposed by the infratemporal fossa approach. Involved dura is resected, and the intracranial portion is removed from the subarachnoid space. Actual invasion of the cerebellum or brain stem by the tumor is extremely uncommon; however, if present, it should be approached by a neurosurgeon.

If the dura has not been violated, closure is uncomplicated. Autograft fat or muscle should be placed into the wound to obliterate the large amount of dead space. A suction drain is placed in order to remove blood and transudated fluid from the large, raw undersurface of the flap. The incision should be closed in layers after closure of the external auditory canal from within.

Closure is most difficult after resection of dura, and problems related to wound closure may dominate the postoperative recovery period. Water-tight dura closure is the ideal. It may be accomplished by suturing or by gluing autograft or homograft dura or fascia to the margins of the dura defect. If a water-tight dural closure is not possible, fat strips may be placed into the wound and dural defect. The sternocleidomastoid and/or temporalis muscle may then be rotated into the defect over the fat strips and sutured into position. Care must be taken if the sternocleidomastoid muscle is used, because removal of its attachment (the mastoid tip) should be presumed to have compromised the superior blood supply. Rotation of the temporalis muscle will be greatly facilitated by removal of the posterior portion of the zygomatic arch.

If the subarachnoid space has been opened, many surgeons avoid the use of a suction drain for fear it will promote CSF accumulation by "sucking" CSF from the subarachnoid space into the wound. We believe that the placement of a shunt from the subarachnoid space into the right atrium reduces CSF accumulation and leakage by controlling CSF pressure. The proximal portion of the shunt tubing is placed into the lateral cerebellar cistern. The wound is carefully closed in layers, and a large pressure dressing is applied.

Individual cases present specific additional problems. Sacrifice of cranial nerves should be dealt with as discussed above. If the internal carotid artery has been bypassed, sacrificed, or even extensively manipulated, then anticoagulation may be appropriate. If bone removal has included the occipital condyle and large portion of C_1, the consideration must be given to stability of the cervical spine. Those patients who have had a portion of the mandible resected should be placed into intermaxillary fixation in the postoperative period.

Approaches to the Medial Skull Base

A variety of approaches to the medial surface of the temporal bone, cerebellopontine angle, and petrous apex are available. They are used most frequently in removing primary intracranial tumors, such as acoustic neuromas and meningiomas. One or more of these approaches may be combined with the approaches to the lateral skull base, described previously, in order to achieve complete tumor removal, especially when there is coexisting intracranial disease.

Retrolabyrinthine Approach

The retrolabyrinthine approach is the one most frequently called upon in removing the intracranial portions of glomus tumors. These tumors most commonly penetrate the dura between the sigmoid sinus and the hard bone of the labyrinthine capsule. The retrolabyrinthine approach provides the most direct access to the intracranial component. It is also useful in dealing with the intracranial portions of schwannomas arising from the jugular foramen. The approach is described in detail in Chapter 50 of this volume; briefly, it consists essentially of opening the posterior fossa dura anterior to the sigmoid sinus.

Translabyrinthine Approach

The translabyrinthine approach is described in detail in Chapter 54 of this volume. Tumors that invade the labyrinth or that dwell on the posterior aspect of the petrous pyramid may be removed after labyrinthectomy. Both the petrous apex and the internal auditory canal are fully exposed. Hearing, if present preoperatively, must, of course, be sacrificed. All residual labyrinthine function on the operative side will be ablated, which may augment difficulties in postoperative recovery.

This approach is especially useful in tumors that involve the internal auditory canal, which is widely exposed after removal of the labyrinth. Excellent exposure of the cerebellopontine angle is achieved at the same time.

Middle Fossa Approach

Some tumors will have proportions that extend into the anterior surface of the petrous pyramid or that involve the floor of the middle cranial fossa. These portions of the tumor can be removed by adding a middle cranial fossa approach to the total operative procedure. (The

approach is described in detail in Chapter 54 of this volume.) The middle cranial fossa approach is useful not only for large, medially placed lesions in the area of Meckel's cave but also for more laterally placed lesions that extend superiorly through the tegmen tympani or roof of the eustachian tube.

Transcochlear Approach

By removing the cochlea, the widest exposure of the petrous apex is achieved. Meckel's cave, the medial portion of the carotid artery in the "siphon", and the cavernous sinus may be approached. It is possible to extend the dissection anteromedially so far as to enter the sphenoid sinus from behind. Congenital cholesteatomas and meningiomas of the middle fossa dura most frequently call for this approach. Hearing and labyrinthine function are, of course, sacrificed.

The transcochlear portion of the procedure is begun after labyrinthectomy, with exposure of the IAC. The facial nerve is fully mobilized and re-routed posteriorly. The three turns of the cochlea are removed with a diamond bur, which controls bleeding, until the carotid artery is fully exposed up to its intercavernous portion. The dura between the superior petrosal sinus and the carotid artery is opened, exposing the clivus as far as the contralateral IAC. The vertebral and basilar arteries, as well as cranial nerve V, are well exposed. Closure follows the principles given for other transdural procedures.

Summary

Tumors of the skull base arise from a variety of anatomic positions within the temporal bone. Each tumor extends itself in a slightly different manner and involves different portions of the cranial base. A large number of vital neural and vascular structures pass through the cranial base, often via complex and circuitous routes. Neoplasms in this area are usually benign. Therefore, en bloc resection techniques that require wide surgical margins and the sacrifice of important structures are entirely inappropriate. Incomplete removal, however, will invariably result in recurrence of disease and significant morbidity and mortality for the patient.

The great challenge facing the skull base surgeon is to remove completely these large, complex lesions while sparing as many important structures as possible. The key to successful surgical removal is flexibility. The operating surgeon must feel comfortable with every approach to and through the cranial base and must be able to combine multiple approaches in a single procedure. Thus, each surgical procedure is "custom-tailored" to each tumor. Without such flexibility and individualization, either important structures will be unnecessarily sacrificed or tumor extirpation will be incomplete.