**Paparella: Volume II: Otology and Neuro-Otology**

**Section 3: Diseases of the Ear**

**Part 2: External Ear**

# Chapter 23: Cysts and Tumors of the External Ear

**Joseph R. DiBartolomeo, Michael M. Paparella, William L. Meyerhoff**

Two embryologic germ layers contribute to the various components that constitute the external ear. Ectodermal derivatives include the epithelial elements and appendages of the skin covering the auricle and external auditory canal. Mesodermal primordial tissues develop into the vestigial muscles of the auricle and the supporting cartilage of the pinna and lateral two-thirds of the external auditory meatus. Congenital aberrations or acquired benign and malignant tumors may arise within any of these tissues. Additional lesions may occur following excessive solar radiation, prolonged hypothermia, chronic irritation, or infection. Less common are the cysts and lesions due to congenital aberrations in the development of the external ear from the first branchial groove between the first and second branchial arches.

## General Classification

### Cysts

**True Cysts.** These include cysts associated with preauricular fistulae and acquired retention cysts. Sebaceous cysts are the most common form of retention cyst, but often an implantation cyst may occur in a post-operative incision.

**Pseudocysts.** Pseudocysts, such as a cystic hematoma, may occur following trauma or within a degenerating tumor.

### Tumors

**Epithelial Tumors.** Benign papilloma or keratoacanthoma may arise from the epithelial elements. The glandular appendages may be the source of cerumenomas or adenomas. The malignant epithelial tumors most commonly seen are squamous cell carcinoma and basal cell carcinoma.

**Mesenchymal Tumors.** Benign mesenchymal tumors include the very common exostoses of the bony external auditory canal, chondroma, myxoma, chondrodermatitis nodularis chronica helicis, hemangioma, keloid, myoma, lipoma, and fibroma. The most significant mesenchymal malignancy is sarcoma.

**Melanocytic Tumors.** Benign nevi or melanoma may occur on the auricle.

**Pseudotumors.** Anatomic variants such as Darwin's tubercle may occasionally be misdiagnosed as a pathologic lesion.

**Keloid Tumors.** These may occur in response to soft tissue trauma or post-surgery.

## Benign Lesions and Abnormalities

### Skin and Soft Tissue Cysts and Tumors

**Pseudocyst.** The collection of fluid into a tissue space without an epithelial lining may be termed a pseudocyst. The swelling may represent a seroma or hematoma following trauma or the cystic degeneration of a tumor. This is most common on the auricle. Treatment is directed toward the underlying cause. When drainage is necessary, the incision should be made on the posterior aspect of the auricle and compression dressings should be applied to the anterior aspect of the auricle to prevent reaccumulation of fluid and to maintain the natural contour.

**Sebaceous Cyst.** Retention cysts of sebaceous glands are common lesions around the auricle, especially in the postauricular sulcus and lobule region. Some may arise from the hair follicle (pilar cysts). They are usually soft and fairly mobile and occasionally there is a definable cyst apex. Symptoms are usually absent unless the cyst becomes very large or infected. Removal may be indicated for cosmetic reasons, because of secondary infection, or if malignant degeneration is suspected. Simple incision and drainage is invariably followed by recurrence. Successful treatment requires surgical excision with the capsule intact.

**Preauricular Cyst and Fistula.** Preauricular cysts are congenital abnormalities resulting from faulty developmental closure of the hillocks of His (first and second branchial arch derivatives). They may be unilateral or bilateral and appear as a pitlike depression in front of the helix or above the tragus. Occasionally the depression may lead to a cyst or epidermis-lined fistulous tract with intermittent or continuous scant, foul discharge. Occasionally one finds familial predisposition, and the cysts may occur in a syndrome associated with sensorineural deafness. Treatment is indicated if the cyst becomes symptomatic or unsightly. Complete surgical removal of both the cyst and its lined fistulous tract is necessary to prevent recurrence and reinfection. (See Part 2, Chapter 20). Preauricular, and rarely postauricular, skin tags with or without cartilage are also the result of faulty fusion of the hillocks of His. They may be removed for cosmetic reasons.

**Dermoid Cyst.** These congenital cysts are a form of teratoma composed of a fibrous wall lined with stratified squamous epithelium and containing hair follicles, sweat glands, and sebaceous glands. They usually present as a round, spongy growth behind the pinna, over the upper part of the mastoid process. There are two theories regarding the etiology of teratoma. One theory alludes to an embryologic ontologic misplacement of cells, suggesting that teratoma represents intrauterine displacement of normal cells that have escaped the influence of the primary organizer. The second theory proposes entrapment of normal tissue by improper fusion of tissues during development. Dermoid cysts can be removed for cosmetic purposes.

**Incisional Cyst or Epidermal Implantation Cyst.** This lesion frequently occurs following incision or trauma in the region of the ear as a result of implantation of squamous epithelium into subcutaneous tissue. Secretions filling the cyst are the products of the secretory cells of implanted sweat or sebaceous glands. Symptoms are usually absent unless

the cyst becomes large enough to produce pressure or unless secondary infection occurs. Indications for removal are cosmetic or when doubt exists concerning the benign nature of the lesion. Surgical removal is the only satisfactory form of treatment.

**Ceruminoma (Hidranoma, Sweat Gland Tumor of the Ear Canal).** The ceruminous glands are modified apocrine sweat glands, present only in the skin overlying the cartilaginous meatus. A ceruminoma is an adenoma of sweat gland origin. Such tumors are rare in humans but common in dogs and cats. This lesion appears as a smooth, intraverted, polypoid swelling in the outer end of the meatus. The presenting symptom is usually a blocked feeling in the ear with no history of otorrhea or previous ear problems. Continued tumor growth may lead to an increasing sensation of pressure. Even benign lesions tend to recur and should be widely excised. Because of the propensity for malignant change or local recurrence, scheduled periodic follow-up is essential.

## Keratosis Obturans

**Cholesteatoma.** These lesions are discussed together because they probably represent the same clinical entity. In this rare condition, a keratotic mass of desquamating squamous epithelium is found in the bony portion of the external auditory canal. The etiology is uncertain but is probably related to faulty migration of squamous epithelial cells from the surface of the tympanic membrane and the adjacent canal, which allows a mass of squamous epithelium and debris to accumulate and intermix with cerumen. The mass appears pearly white and glistening, as does cholesteatoma of the middle ear. For this reason keratosis obturans is often called cholesteatoma of the external auditory canal. Keratosis obturans is frequently associated with bronchiectasis and sinusitis in younger patients. Pain is the common presenting symptom and results from erosion of the osseous meatus. Conductive hearing loss and otorrhea may be present. Treatment consists of periodic removal of accumulated debris, which can be difficult and painful for the patient, occasionally requiring general anesthesia. Topical therapy for associated external otitis should be employed. For obstinate cases in which prolonged conservative measures fail, canaloplasty may be successfully employed.

**Keloid.** Keloids especially occur in the lobules of pierced ears. Because of social customs, the incidence is greater in females. Keloids may occur spontaneously following surgical procedures in black patients or following secondary infection in other races. It also may occur following surgical or radiation therapy for carcinoma of the auricle. Keloid is a benign connective tissue hypertrophy characterized by a smooth, pink, rounded scarlike tumor that invariably follows trauma or incision in the skin. It appears to be secondary to a defect in collagenase, which results in the overgrowth of collagen as opposed to a hypertrophic scar composed of immature collagen that has failed to convert from the tertiary to the quaternary form. Complete eradication is difficult because of the tendency for recurrence. The traditional treatment has been surgical removal, followed by immediate postoperative low-dose (500 rads) irradiation. Occasionally, injection of Kenalog at the time of surgical excision is helpful. Recent reports indicate successful treatment using the $CO_2$ laser.

**Papilloma or Verrucous Wart.** The etiology of the papilloma or verrucous wart is generally considered to be a virus that provokes local hypertrophy of the papillae. They may present as flat gray plaques but more likely appear as cauliflower outgrowths of the pinna.

When they occur in the external auditory canal, papillomas must be differentiated from polyps or a malignant growth. Papillomas are rare in the external auditory canal and even less common on the auricle. The treatment is wide excision or cryosurgery.

**Seborrheic Keratosis.** Seborrheic keratosis (senile wart) is a proliferation of basal cells with the formation of epithelial cysts and hyperkeratosis. Melanocytes and melanin pigment may be present, imparting a yellow, light brown, gray, or black color. They usually are elevated, irregular, and greasy, but many appear flat and dry. Small pieces may flake off, with minor trauma, and subsequent slight bleeding may occur. These growths present during middle age, becoming the most common cutaneous tumors of the older age groups. They are almost always benign but should be excised if the diagnosis is in question or if they become cosmetically unpleasant.

**Senile Keratosis.** This is the most common precancerous cutaneous lesion and is the result of excessive solar exposure. Although usually asymptomatic, these lesions may cause discomfort or a pressure sensation. They vary in size (rarely greater than 1 centimeter in diameter) and may be yellow, brown, or black. They lack the characteristic pits and furrows of seborrheic keratosis (a similar lesion without malignant potential). Treatment is excision or the application of topical 5-fluorouracil.

**Keratoacanthoma.** This benign lesion is of clinical significance because of its physical resemblance to squamous cell carcinoma. However, the clinical picture consists of rapid growth of a lesion to 1 centimeter or more in diameter within a month to 6 weeks. A central keratin core or plug forms but undergoes spontaneous resolution over several weeks. They most commonly occur as solitary lesions with a predilection for sun-exposed areas. Single lesions can be excised, but multiple lesions should be histologically reviewed in detail to rule out the possibility of squamous cell carcinoma. Multiple lesions may occur in a familiar basis, usually do not involute spontaneously, but may be treated successfully with retinoic acid.

**Cutaneous Horns.** These benign projections of unusually cohesive keratin may form on epidermal nevi, warts, sebaceous keratosis, precancerous keratoses, or squamous cell carcinoma, making total excision with histologic examination the treatment of choice.

**Nevi.** Benign nevi are fairly common on the external ear and are usually of the intradermal type. They frequently are congenital, benign proliferative lesions of the skin and occasionally of mucosa, which are presumably hereditary. The *blue nevus* is a focal, pigmented lesion that is slightly elevated and usually less than 0.5 centimeters in diameter, with a regular outline. Histologically, these nevi are composed of closely grouped melanocytes and melanophages located in the mid-dermis. Surgical excision may be indicated for cosmesis. *Moles* are hyperpigmented skin lesions located at the junction of the dermis and epidermis (junctional), in the dermis (dermal), or both at the junction and in the adjacent dermis (compound). They may be removed for cosmetic reasons, but prophylactic removal for prevention of degeneration into malignant melanoma is probably unjustified. *Melanoma* is the result of malignant transformation in any melanocyte, and its apparent predisposition for moles is probably based on the higher concentration of melanocytes within the mole rather than on the malignant potential of the mole melanocyte itself.

**Nevus Sebaceous of Jadassohn.** This is a sebaceous hamartoma derived from the skin and its glandular adnexa. It is usually observed shortly after birth and appears as a solitary, irregular, yellow-to-amber, oily flat lesion. It may be seen in association with mental retardation and is considered potentially malignant. Early excision is recommended.

**Lipoma.** Benign fatty tumors, which are very common in other areas of the body, are extremely rare in the external ear. They are seen in adults and occur usually in the postauricular region or lobule. Lipomas are usually symptomless and present as soft, freely mobile, slow-growing tumors. Indications for surgical excision may be cosmetic or diagnostic.

**Xanthoma.** Although not a true neoplasm, a xanthoma can resemble a tumor. These lesions occur in disease states associated with a disturbance of cholesterol metabolism, and they consist of cholesterol fatty acid esters in connective tissue and epithelial cells. Xanthomas may develop on the auricle or in the canal. They have a characteristic yellowish appearance and are relatively soft in consistency. Numerous lesions almost always exist elsewhere. The treatment, if necessary, is electrocauterization for small lesions and surgical excision for large nodular ones.

**Myomas and Fibromas.** Both of these are extremely rare. Myomas of the auricle area are usually composed of striated muscle, referred to as rhabdomyomas, and arise from the extrinsic muscle of the ear. Myomas composed of smooth muscle are called leiomyomas and can arise from the arrectores pilorum muscle or from the muscular wall of blood vessels. Fibromas are usually firm or nodular subcutaneous swellings. The diagnosis is by biopsy, and treatment is surgical excision

**Myxoma.** A myxoma is a tumor composed of mucin-forming connective tissue. Myxomatous degeneration can occur in fibromas and lipomas and should not be confused with a true myxoma. These tumors are quite rare in the vicinity of the external ear. They usually occur in the subcutaneous tissues and are soft, fluctuant, circumscribed, and translucent. Fibromas are usually firm lesions composed of white fibrous tissue and a scant supply of blood vessels. The diagnosis is established by biopsy, and treatment is surgical excision.

**Mixed Tumors of Salivary Gland Type.** Mixed tumors of salivary gland type may develop in the skin of the external auditory canal, although this is rare. Such tumors have been found in many parts of the body not associated with either major or accessory salivary glands. There is little doubt that these are of ectodermal origin, but the mechanism and timing of the ectodermal implantation and development remain to be determined. These neoplasms present as movable masses in close approximation to the overlying skin. The treatment is local excision and the prognosis is excellent.

**Adenoma.** Adenomas are rare tumors composed of glandular structures. They probably originate from sebaceous glands in the fibrocartilaginous portion of the external auditory canal. These tumors are usually small, soft painless swellings, located at the entrance of the meatus and are asymptomatic unless large enough to cause obstruction of the external auditory canal, with a resultant conductive hearing loss and interference with the exteriorization of wax. Secondary external otitis frequently develops. Treatment is complete surgical excision followed by electrocauterization of the tumor bed. The diagnosis is made

by histologic examination.

## Vascular Tumors

**Congenital Tumors.** Two types of congenital tumors may involve the auricle in children: angioma (hemangioma and lymphangioma) and dermoid tumors.

**Strawberry Mark.** This type of immature angioma, also called nevus vasculosus, is present at birth but may not become apparent until 3 months of age. The lesion grows during childhood but usually regresses after puberty. Conservative treatment consists of periodic observation as most capillary hemangiomas regress with time. Rarely is surgical or laser excision indicated unless the lesion obstructs the external auditory canal resulting in a conductive hearing loss or if hemorrhage occurs because of repeated abrasion.

**Cavernous Hemangiomas.** This lesion is usually an irregular, soft, bright-red or deep purple, papular nodular mass that is easily compressible. It increases in size when the patient strains or there is an increase in intracranial pressure. Some lesions are associated with an occult internal hemangioma or spinal cord arteriovenous malformations. When both superficial and deep elements are present, the lesion is referred to as a *mixed hemangioma.* For persistent lesions or those that tend to bleed or ulcerate, reconstructive or laser surgery may be indicated.

**Compact Hemangioma.** This is an encapsulated growth consisting of firm alveolar masses within the subcutaneous tissues. This rare lesion is best treated by excision.

**Nevus Flammeus.** This is not actually a nevus but a vascular dilatation most frequently called "port wine stain" because of its clinical appearance. It is formed by a diffuse plexus of mature vessels in the dermis. As the patient grows, the lesion does not increase in size except in relation to the growth of the part affected. Port wine stain may also be associated with an encephalo-trigeminal angiomatosis or may be transmitted as an autosomal dominant trait and variant of the Sturge-Weber syndrome.

**Lymphangioma.** Lymphangiomas on the pinna are rare. They are similar to hemangiomas except that the channels contain lymph instead of blood. Accepted treatment is surgical excision. One must remember that treatment of the patient with a congenital lesion should always involve counseling of the parents to address the denial or guilt that parents may have regarding a congenital lesion.

## Lesions of the Cartilaginous External Ear

**Darwin's Tubercle.** This is an atavistic lesion, or genetic marker, occasionally misdiagnosed as a calcification of the auricle or cartilaginous deformity. It is homologous to the tip of the mammalian ear. It usually appears as a 2-millimeter elevation on the posterior lateral rim of the auricle at the junction of the upper and middle third of the helix. It is completely painless with digital compression.

**Chondrodermatitis Nodularis Chronica Helicis.** In some countries this condition is known as *Winkler's disease.* These small troublesome growths are approximately the size of

a small pea, occur on the free edge of the pinna, and appear as firm elevated nodular lesions with a grayish crust on the surface. Exquisite tenderness with digital compression, out of proportion to its size, differentiates these nodules from a wart or epithelioma. They are much more common in males than females, and their cause is unknown but may be related to chilling. They must be differentiated from basal cell carcinoma and the tophaceous deposit of gout. Relief of pain may be obtained by the injection of hydrocortisone acetate (25 mg per mm of size). Later, the lesion may be excised if necessary.

**Rheumatoid Arthritis.** Cutaneous and subcutaneous nodules of rheumatoid arthritis may appear on the auricle. These nodules are frequently painful, with necrotic centers. Treatment is directed toward the systemic process.

**Gout.** Gout is characterized by hyperuricemia, recurrent painful arthritis responsive to colchicine, and deposition of urates in nonarticular tissues. Asymptomatic urate deposits may be found in the helix and antihelix; they vary from one millimeter to several centimeters in size. The lesions usually contain monosodium urate needle-like crystals. They are distinguished by appearance and history from rheumatoid nodules, Darwinian tubercules, xanthoma tuberosum, and chondrodermatitis nodularis chronica helicis. *Tophaceous deposits* of gout are highly specific lesions that occur most commonly on the helix and may be confused with rheumatoid nodules. They are painful, salmon pink-subcutaneous nodules that occasionally drain a chalky white material consisting of monosodium urate crystals. Like rheumatoid arthritis, treatment is directed toward the systemic disorder.

**Chondroma.** This is a rare lesion arising from the cartilage of the auricle. It is usually associated with trauma to the external ear. *Enchondromas* are cartilaginous spicules occurring in the external canal; they may cause obstruction. Treatment is complete surgical excision because of their predilection for continued growth and eventual deformity of the auricle. If excision is not complete, the portion that remains will continue to grow.

**Calcinosis.** Calcification may occur in auricular cartilage and is often asymptomatic until it is discovered fortuitously. It may occur as a result of the process of dystrophic (primary) or metastatic (secondary) calcification. Primary calcification occurs when deposition takes place in damaged, or necrotic tissue in patients whose serum calcium and phosphorus levels are normal. Frostbite and mechanical trauma are the most common causes of primary or dystrophic calcification. Secondary calcification occurs in association with a disturbance of calcium metabolism. It may be seen in association with hypercalcemia, vitamin D intoxication, sarcoidosis, milk-alkali syndrome, primary or secondary hyperparathyroidism and in tumoral calcinosis associated with hyperphosphatemia.

**Ossification of the Auricle.** Ectopic ossification has been reported in several structures of the head and neck, including the inner ear and tracheal cartilages. It may also affect the auricle. It may be clinically distinguished from calcification of the auricle by the presence of a trabecular pattern detected on a radiograph of the auricle. The diagnosis is irrefutable only by histologic analysis.

## Tumors of the Osseous External Auditory Meatus

### Hyperostosis

**Osteoma.** Proliferation of cortical bone in the external auditory canal may occur in two forms: exostoses (multiple compact hyperostoses) and the single osteoma.

**Exostoses.** Multiple exostoses are the most commonly encountered tumors of the external auditory canal. They rarely occur during childhood and are much more common in males than females; they usually are bilateral. The etiologic factor most likely responsible for the formation of exostoses is prolonged and repeated stimulation of the external auditory canal of susceptible individuals by cold water; this is most commonly seen in those who swim in salt water. Other factors that may play a role in the formation of exostoses include chronic irritation from infection, eczema, and trauma. The first response of the bony ear canal to hypothermia is a change in the configuration of the oval bony external auditory canal lumen to a V-shaped configuration referred to as "Di's notch". In time, the exostoses enlarge as smooth rounded nodules of extremely hard bone covered with normal skin and attached by a broad base usually at the suture lines. Smaller rounded nodules usually occur on the superior bony wall adjacent to the annulus. Usually they are asymptomatic and require no treatment until they become large enough to cause obstruction, impair hearing, or foster the retention of cerumen and debris. This leads to fullness or pressure in the ear, otorrhea, and otalgia. When exostoses become symptomatic, surgical excision may be advisable. It has been documented that the disease is preventable if the perfusion of the bony ear canal by cold water is prevented. This can be accomplished if surfers and swimmers use vented ear plugs. Divers should wear a hood instead of ear plugs.

**Osteoma.** Single, cancellous osteoma, unlike exostosis, is a relatively rare lesion of the external auditory canal. Osteoma invariably occurs unilaterally and may resemble a foreign body or cyst. Conductive hearing loss and discomfort are common symptoms. If not treated, the solitary cancellous osteoma will eventually occlude the meatus. For diagnostic purposes it is usually possible to encircle this growth with a bit of cotton tightly wound about a wire applicator or other instrument. Palpation reveals a hard, bony consistency. The size may vary from that of a small millet seed to over 2 centimeters in diameter. Surgical removal should be advised in every case; otherwise, the cancellous osteoma will continue to enlarge, causing occlusion of the canal and secondary infection.

### Malignant Tumors

We have divided malignant tumors of the external ear into two groups because of the marked difference in the clinical picture and mode of management: (1) those involving the auricle, and (2) those involving the external auditory canal, which includes lesions that occur within 1 centimeter of the external auditory canal.

### Incidence

Malignant lesions of the auricle are much more common than those in the external auditory canal or middle ear. Cancer of the pinna accounts for approximately 4 to 8 per cent of all skin cancers. Bezold reported only four malignant tumors of the external auditory canal

in 20.000 otologic cases. Of 6605 cases of external ear disease studied at the Royal Infirmary of Edinburgh, Scotland, only 13 proved to be malignant. Only 28 patients with malignant tumors of the external auditory canal were encountered out of 6500 general admissions studied in a New York Hospital.

In the clinic of one of the authors, a study of 32 cases of temporal bone malignancy, plus 10 pathologic cases of temporal bone cancer, demonstrated the difficulty of assessing the incidence of this form of malignant disease. In many of these patients, the primary cancer probably occurred in the ear canal, thereby suggesting a more common occurrence for such lesions than is generally considered. After invasion of the middle ear and mastoid occurs, however, it is difficult to determine the site of primary origin. This emphasizes the importance of early diagnosis.

In general, about 80 to 85 per cent of aural cancers involve the auricle, 10 to 15 per cent the external auditory meatus, and 5 to 10 per cent the middle ear and mastoid.

## Malignant Tumors of the Auricle

Approximately 90 per cent of all skin cancers occur in the head and neck region, and 6 per cent of all skin cancers occur on the ear. Although the etiology of carcinoma of the auricle is unknown, various factors have been incriminated. These factors are local and include chronic trauma from sun and wind, chronic infection, chronic irritation, chronic eczema, psoriasis, and chemical and x-ray injury. Squamous cell carcinoma is the most common malignant tumor of the auricle, followed by basal cell carcinoma. Other, rare malignant tumors that may occur are adenocarcinoma, cylindroma, and melanoma. The majority of tumors occur on the helix and anthelix, with a diminishing incidence on the concha, lobule, tragus, and antitragus.

**Squamous Cell Carcinoma.** Squamous cell carcinoma occurs six times more commonly on the auricle than in the canal and, because of easy visibility, earlier recognition and diagnosis are more likely. It usually occurs in older men on the posterosuperior portion of the pinna. In women, the concha close to the external auditory meatus is frequently involved. Auricular cancer grows extremely slowly, and metastasis to regional lymph glands develops late. The earliest change is a thickening of the skin associated with scaling. As the cancer grows, a firm, painless, pale outgrowth develops. Eventually the surface disrupts, with the formation of an ulcer with a raised edge. Biopsy should be made of any lesion for which the clinical diagnosis is in doubt.

The treatment of choice for squamous cell carcinoma of the auricle is wide local excision. If the lesion occurs on the helix or anthelix, a wide excision that includes 1 centimeter or more of normal tissue should be made. If there is uncertainty in obtaining an adequately safe margin for excision, the chemosurgical techniques as described by Mohs (1947), which is also applicable for basal cell carcinoma, can be employed. In brief, this method calls for painting the lesion with zinc chloride, followed by frozen sections for serial assessments of an adequate excisional margin. For large, fungating carcinomas, or those involving the concha, the entire auricle must be sacrificed. More conservative procedures are to be used reservedly and only for small, localized growths. If regional lymph nodes are involved, a radical en bloc dissection may be necessary. Radiotherapy may be used in

combination with surgery or for recurrence. The prognosis for squamous cell carcinoma of the auricle depends on the site and extent of the lesion as well as the mode of treatment. The long-term results of surgical excision are superior to results of treatment by radiation therapy. Small lesions located on the helix or anthelix, which lend themselves to complete wide excision, have a 5-year cure rate of 95 per cent, whereas those lesions near the cartilaginous meatus have a much poorer prognosis.

**Basal Cell Carcinoma.** Basal cell carcinoma is the result of a proliferation of the basal cells of the epithelium. Although more common on the face than squamous cell carcinoma, it is less common on the auricle. Men are affected more often than women. Onset is usually later in life than squamous cell carcinoma, occurring most frequently in the fifth or sixth decade, and etiology is unknown. The tumor is very slow-growing and usually does not metastasize, but it is locally aggressive and may destroy cartilage and bone. It is usually asymptomatic, and the diagnosis is made by biopsy. Basal cell cancer can frequently be differentiated from squamous cell cancer by its clinical appearance. Whereas squamous cell carcinoma is an outgrowth, basal cell carcinoma develops as an ingrowth. First, a flat, painless, slightly raised lesion appears, followed by development of a rolled edge with a penetrating ulcer which bleeds readily. This "rodent ulcer" progresses by circumferential and deep growth. The treatment of choice is wide surgical excision, and the prognosis is excellent in those patients in whom the lesion was small enough to be wholly excised. Chemosurgery, as described for squamous cell carcinoma, may also be considered, and radiotherapy should be contemplated if the lesion is extensive. The combination of surgery plus full-dose radiation often results in further breakdown in bone and soft tissue. When the tumor has penetrated beyond the reach of surgery, the prognosis is poor.

**Hutchinson's Melanotic Freckle.** This lesion develops from melanocytes and may develop into a malignant melanoma. It represents a proliferation of typical melanocytes in the junctional zone. The pigment distribution is irregular, and there is severe solar degeneration in the dermis. The lesions usually begin as small, brown macules on the face in the antitragal area. As they grow, the coloration becomes uneven and the lesion develops a papular form. When malignant degeneration occurs, the prognosis is somewhat better than that for other malignant melanomas.

**Malignant Melanoma.** Malignant melanoma of the external ear, unlike the benign nevus, is fortunately an extremely rare occurrence. The growth may develop from a small, rounded, darkly pigmented tumor of "nevus" or from any melanocyte, as described earlier. When the lesion increases rapidly in size, the patient may seek consultation with an otolaryngologist. Pain is not a usual symptom until either ulceration occurs or nerve structures are involved by direct extension. If a long-existent pigmented mole suddenly becomes elevated, increasingly pigmented, or ulcerated or bleeds, and is attended by localized discomfort or pain, it should be regarded as a malignant melanoma until proven otherwise. Excisional biopsy will reduce the chance for dissemination of the malignant cells by cutting across the tumor and providing tissue for histologic diagnosis. Malignant melanoma behaves unpredictably, thus requiring a guarded prognosis. The lesion may have an innocuous appearance, yet early widespread metastasis may already have developed.

**Miscellaneous Tumors.** Other, less common tumors can occur in the auricle, such as cylindroma (or cystic adenoid epithelioma), adenocarcinoma, and sarcoma. They are

considered in the next section with tumors of the external auditory canal.

## Malignant Tumors of the External Auditory Canal

Since there is a good likelihood that many, if not most, cancers of the middle ear and mastoid originate primarily in the external auditory canal, it is probably true that this lesion is far more common than is currently recognized. It is quite imperative, therefore, to emphasize the importance of early diagnosis of external auditory canal lesions. As with carcinoma occurring elsewhere in the body, an early diagnosis with adequate treatment is necessary if good results are to be expected.

Since the various malignant tumors of the canal usually present with similar symptoms and findings, they are discussed together. Common symptoms include pain, otorrhea, bleeding, fullness, and decreased hearing. Pain can be a very important feature; it usually occurs in an early stage of the disease and may be very intense, seeming to be out of proportion to the physical findings. Any patient presenting with otalgia should have microscopic otoscopy and palpation for any evidence of tenderness in the canal. If there is a suspicious lesion, biopsy should be done without delay. Persistent external otitis, especially when localized, may also warrant a biopsy. If the lesion is exfoliative and small, an excisional biopsy is best. Pain may result from associated infection, pressure, or invasion of the perichondrium and periosteum. Pain has been reported as an early occurrence in adenoid cystic carcinoma and is correlated with histologic evidence of early perineural infiltration by the tumor. Another important symptom is otorrhea or bleeding, which is usually a late finding. This may occur in association with pain. The most common finding is an aural growth which, at times, is clinically difficult to differentiate from an aural polyp, granulation tissue, and external otitis. It is important to stress here that if there is any doubt, a biopsy should be taken and sent to a laboratory for microscopic examination.

In the treatment of carcinoma of the external auditory canal it is important, whenever possible, to distinguish the exact location of tumor origin. Metastasis to regional lymph glands is slow to occur, and the first group of nodes usually involved is the pretragal group, the superficial and deep cervical nodes being involved later. Anterior canal lesions spread through the lymphatics in the fissures of Santorini, and pretragal involvement can be assumed. Widespread distant metastases are rarely seen. If the primary lesion is unchecked, death occurs from local extension to other structures before metastases occur.

Treatment consists of wide surgical excision, if possible. Adequate surgery includes (1) removal of the drumhead, anterior canal wall, condyle, and parotid gland for isolated anterior canal regions; (2) radical mastoidectomy as a minimum for posterior canal lesions; and (3) consideration of total removal of the temporal bone when there is middle ear or mastoid involvement. Combined radiation therapy, although occasionally indicated in these patients, may lead to devitalization, sequestration of bone, breakdown of soft tissue, and drainage, rendering healing and identification of possible recurrences difficult.

**Basal Cell Carcinoma.** Basal cell carcinoma in the external auditory canal is much less common than on the auricle. Symptoms are as described previously. Whereas basal cell carcinoma of the auricle has a relatively good prognosis, especially if found early, basal cell carcinoma of the external auditory canal has a much poorer prognosis. This is most likely due

to the fact that it is usually discovered later in the course of the disease, and bony erosion or invasion of the middle ear may already have occurred.

**Adenocarcinoma.** Adenocarcinoma is probably the least common malignant tumor encountered in the canal. These lesions usually originate in the sweat or sebaceous glands. The management of adenocarcinoma of the external ear is similar to that of other malignant tumors in this region.

**Adenoid Cystic Carcinoma.** Adenoid cystic carcinoma of the external auditory canal is not common. It is an adenocarcinoma usually arising from the ceruminous glands and is frequently called cylindroma. The average age at onset of symptoms is in the fifth decade of life, but symptoms may occur at an earlier age. The tumor is characterized by its slow growth, and the symptoms may last for many years. The most outstanding symptom is pain, which usually occurs early and, as the disease progresses, becomes extremely severe and persistent. It usually starts as a sharp pain and may extend to the temporal and postauricular regions, and is the result of this tumor's predilection for nerve invasion. As the disease progresses by local extension, considerable bony destruction occurs. Clinically these tumors appear as yellowish, firm, smooth masses in the canal with numerous small, dilated blood vessels over the surface. Distant metastases and local recurrences are common, with the lungs and kidney being the usual sites. Metastasis occurs late in the course of the disease, usually 10 or 15 years after the onset of symptoms. Radiation therapy alone has little to offer, and surgery offers the best chance.

**Sarcoma.** Sarcoma is a malignant tumor of mesoblastic derivation composed chiefly of rapidly multiplying cells resembling those of connective tissue. Many types of sarcoma may occur in the external ear, including chondrosarcoma, fibrosarcoma, osteosarcoma, myxosarcoma, lymphosarcoma, and the undifferentiated cylindrical cell type of sarcoma. However, all these are extremely rare and are considered here as one group. As with other malignancies, the etiology of these tumors is unknown, although trauma has long been regarded as a possible causative factor. Sarcoma frequently affects younger individuals than does carcinoma. Sarcomas usually present as round and nodular swellings; their rapid increase in size may be the first sign that arouses suspicion. If the diagnosis of sarcoma is confirmed, the prognosis will relate to the degree of differentiation and the site of origin.

Excision of well-differentiated sarcoma of the pinna may often result in a permanent cure. An undifferentiated sarcoma in the external auditory canal may lead to death within months because of rapid growth and destruction. In general, however, sarcomas are highly malignant tumors with a tendency to early metastasis. The management of sarcoma is similar to that of carcinoma.