Chapter 22: Epithelial tumours of the external auditory meatus and middle ear

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Case reports of cancer of the ear began to appear about 1875. Furstenberg (19240 stated that carcinoma of the ear was first reported around 1775 by Wilde, Schwartze and others, but this statement is wrong: first because histopathology was not available in 1775; and second because Wilde was not born until 1815 and Schwartze not until 1837! This typographical error was later copied by others and illustrates well the dangers of quoting historical references secondhand, particularly when the original paper is in another language.

The first systematic description of the disease appeared in 1883 in Politzer's classic textbook, based mainly on cases reported by others, for example Schwartze. It was already recognized that carcinoma of the middle ear arises 'either during an existing suppuration of the middle ear or after exhaustion of carious processes in the temporal bone'.

The available literature was reviewed in Zeroni's monograph of 1900, and by Newhart in 1917. In 1921, Broders presented a study of 63 epitheliomata of the ear. His paper covered nine tumours of the external meatus, one of the middle ear and 53 of the auricle. It was the first pathological account of a large number of cases and it includes interesting details of histological types and differentiation, lymph node metastases, etc, but, as all the cases were grouped together, it is not possible to deduce anything specific about tumours at individual sites.

The major contributions in the twentieth century are the accounts of radiotherapy by Lederman (1965) and of surgery by Lewis (1975).

Aetiology

External auditory meatus

The most commonly discussed aetiological factor is chronic inflammation. Irradiation injury in the form of repeated treatment of external otitis has also been mentioned. Highly speculative uninvestigated causes include carcinogens produced by the indigenous microbial flora. Aflatoxin B, a potent hepatic carcinogen, is produced by Aspergillus flavus, an occasional transient contaminant of the ear canal. Equally speculative is the production of carcinogens within cerumen. If chronic inflammation and infection are important it is curious that carcinomata arising in patients with long-standing chronic otitis media always do so in the remnants of the middle ear cleft and never in the external auditory canal over which the discharge also flows.

Tumours of the ear can be chemically induced in animals. One dose of azoxymethane (a derivative of dimethylhydrazine) injected into rats induces squamous cell carcinoma of the sebaceous glands of the external auditory meatus in about 15% of animals. Epidermoid carcinomata develop spontaneously in the external canal and middle ear of elderly female gerbils.
Pre-existing chronic otitis media is commonly believed to be the main predisposing cause of carcinoma of the middle ear.

The progression from chronic otitis media to squamous carcinoma was certainly well known a century ago and was reported by Politzer in his textbook of 1883; a frequent history of chronic otitis media in patients with carcinoma of the middle ear has been reported over a long period. As many as 85% of all cases of malignancy have chronically discharging ears.

Irradiation-induced carcinomata have been recorded after irradiation therapy to the head and neck, and exposure to radium has also been implicated.

Pathological anatomy

External auditory meatus

The external auditory canal can be subdivided into two parts:

(1) A cartilaginous portion: tumours arising here spread easily because the cartilaginous walls present little resistance; spread may be anteriorly into the parotid gland or posteriorly into the postauricular sulcus. The cartilage of the external auditory canal is an inward prolongation of that of the pinna, so that tumours may readily spread in this layer outwards into the concha.

(2) The bony portion: this is surrounded by dense bone which provides an effective barrier to spread of the tumour which is then deflected along the canal into the middle ear.

Middle ear and mastoid

The middle ear and mastoid may be divided into two parts: petromastoid and tympanotubal.

Petromastoid tumours

The petromastoid unit includes the tympanic cavity and the mastoid antrum. Tumours arising here may include:

(1) those limited to the tympanic cavity
(2) those limited to the mastoid antrum
(3) those involving the tympanic cavity and mastoid antrum
(4) those involving the tympanic cavity and external auditory canal.

Descriptions of the pathological anatomy in these cases rest on the descriptions of normal anatomy. Almost all carcinomata of the middle ear arise in patients with long-standing chronic otitis media who have usually undergone previous mastoidectomy. The anatomy is then very different from the normal state and this factor contributes largely to the poor
prognosis in these patients. Superiorly, the mastoid cavity is bounded by the thin tegmen tympani which may have been breached at previous surgery or disease. Medially, the promontory of the middle ear is usually exposed, with two important practical results: first, the facial nerve is either exposed or covered only by a thin layer of bone in its intratympanic course so that facial paralysis is a frequent accompaniment of disease in this area; second, there are several pathways readily available for medial spread of the tumour into the petrous apex. The oval and round windows are theoretical pathways, but more important is the track of cells leading above, below and behind the labyrinth into the petrous apex. The tumour thus gains access to the petrous pyramid lying medial to the bend of the internal carotid artery.

In the patient who has undergone a mastoidectomy operation, the tumour has ready access to the base of the skull, particularly the jugular foramen, and this fact is one explanation for the paralysis of the lower cranial nerves. An alternative explanation is metastasis to the node situated over the transverse process of the atlas in the lateral compartment of the parapharyngeal space.

Post-mortem examination of patients who died of middle ear carcinoma has shown the possible pathways of spread. In addition to local invasion of the remnants of the ossicles, the stapedius muscle and the facial canal, there are two important directions of spread. The first is within the eustachian tube; the medial bony wall of the eustachian tube and the associated bony wall of the middle ear cavity are separated from the carotid canal by a thin layer of bone, and this appears to be a frequent route of spread of tumours to the carotid canal. Second, the tumour spreads into the mastoid air cells, penetrates the thin bony wall of the posterior group of air spaces and thus reaches the internal auditory meatus. The structures in the labyrinth are remarkably resistant to the tumour.

**Tympanotubal tumours**

These tumours either arise in the middle ear and spread into the bony eustachian tube or may even arise within the tube itself. The bony portion of the eustachian tube is anatomically part of the tympanic cavity and has been termed the protympanum. Tumour probably spreads within the surrounding fascial space rather than along the tube. Invasion of these fascial spaces gives the tumour access to the trigeminal or oculomotor nerves in the lateral wall of the cavernous sinus.

**Lymphatic drainage**

The lymphatic drainage of the external auditory meatus follows the same course as that of the auricle, that is it may go in one of three directions:

1. anteriorly to the parotid lymph glands, especially to the gland in front of the tragus
2. inferiorly to the lymph glands that lie along the external jugular vein and those under the sternomastoid muscle
3. posteriorly to the mastoid lymph nodes.

The lymphatics of the middle ear and mastoid are less well defined. Anatomical texts state that the lymph vessels are arranged, like the blood vessels, in two sets on the medial and
lateral surface of the tympanic membrane. However, in virtually all patients with carcinoma of the middle ear, the tympanic membrane has been destroyed. The lymphatic pathways in such patients do not appear to have been defined, but are probably sparse, as is shown by the paucity of lymph node metastases in this disease.

**Tumour types**

The following classification of tumours of the temporal bone is expanded from that of Lewis (1975).

**Benign tumours**

(a) Epithelial: primary cholesteatoma (primary cholesteatoma, choristoma, adenoma)

(b) Mesenchymal
   (i) jugulo-tympanic paraganglioma (glomus jugulare tumour, chemodectoma)
   (ii) osteoma
   (iii) haemangioma
   (iv) neurogenic tumours
   (v) xanthoma
   (vi) giant cell tumour
   (vii) benign osteoblastoma

**Malignant tumours**

(a) Primary
   *Epithelial*
   (i) squamous cell carcinoma
   (ii) adenocarcinoma (hidradenocarcinoma)
   (iii) melanoma
   (iv) basal cell carcinoma
   (v) sebaceous cell carcinoma
   *Mesenchymal*
   (i) sarcoma
   (ii) multiple myeloma
   (iii) haemangioendothelioma
   (iv) malignant xanthoma

(b) Secondary
   *Direct extension from:*
   (i) nasopharynx
   (ii) external ear
   (iii) parotid
   (iv) meningioma
   *Distant metastases from:*
   (i) kidney
   (ii) lung
(iii) prostate  
(iv) breast  
(v) uterus.

Only epithelial tumours arising in the external auditory meatus and middle ear will be discussed here.

**External auditory canal**

**Squamous carcinoma**

Squamous carcinoma constitutes about 90% of all malignant tumours. It can originate in any portion of the external auditory meatus but most often arises in the bony rather than in the cartilaginous portion. Most tumours develop slowly, although occasionally rapid growth is seen.

Invasion of the cartilage of the membranous portion is usually a late development. Since the cartilage provides a barrier, extension is usually along the perichondrium. The tympanic membrane limits spread of the disease, but eventually weakens and breaks down, allowing invasion of the middle ear. Facial nerve paralysis develops when the middle ear and mastoid have been involved. Squamous cell carcinoma extends through the cartilaginous and bony walls of the canal late in the disease, invading the surrounding parotid gland anteriorly or the sternocleidomastoid muscle insertion inferiorly and posteriorly.

Metastasis to cervical lymph nodes from squamous cell carcinoma in this area is a late manifestation of the disease occurring in about 20% of patients. Lesions in the posterior wall of the canal usually metastasize to the nodes in the subcutaneous tissue overlying the insertion of the sternocleidomastoid muscle. Tumours originating in the inferior portion of the canal generally metastasize directly into the subdigastric (jugulodigastric) lymph nodes, while those originating in the anterior portion of the canal metastasize to the preauricular lymph nodes lying in the parotid gland.

**Adenocarcinoma (including hidradenoma)**

The so-called ceruminous glands of the external auditory meatus are typical apocrine sweat glands. Their secretion is a watery fluid devoid of lipids. These glands do not secrete the wax of the meatus which is produced by sebaceous glands, and their title is therefore not justified. Tumours arising from these ceruminous glands may be divided into adenomata, mucoepidermoid carcinoma, adenoid cystic carcinoma and adenocarcinoma.

The terms hidradenoma and ceruminoma are synonymous, and either may be used as a 'blanket' term for all these benign and malignant tumours. Hidradenoma is a better term because ceruminoma is a misnomer as the so-called ceruminous glands, as mentioned above are modified sweat glands.

Hidradenomata have two histological features of diagnostic significance: first, a two-layered epithelial structure analogous to that of the normal sweat gland consisting of an inner oxyphilic columnar layer and an outer myoepithelial layer. The second is a variable degree
of glandular stroma. They may also have a papillary or a cystic pattern. They cause obstruction but seldom pain. An adenoma which is clearly benign requires local excision.

For mucoepidermoid carcinoma, wide excision of the entire external auditory canal, radical mastoidectomy, excision of the mandibular condyle and total parotidectomy with preservation of the facial nerve is recommended. Adenoid cystic carcinoma is by far the most common ceruminous tumour and resembles that found elsewhere. It causes pain, and has a long natural history ranging from 10 to 30 years terminating in death from local invasion or distant metastases. Radiotherapy has little to offer these patients and the recommended treatment is wide excision of the external auditory canal and surrounding bone, part of the pinna, extended radical mastoidectomy, excision of the dura, total parotidectomy, and excision of the mandibular canal and any involved surrounding structures. Finally, simple adenocarcinoma may occur; it has a wide histological spectrum of glandular adenoid tubular and adenoid cystic patterns. The basic pattern is that of adenocarcinoma with two-layered eosinophilic glands. This tumour infiltrates widely into the middle ear and mastoid, etc. It is a very aggressive disease often presenting with facial paralysis, and usually proving fatal within 4 years.

Malignant melanoma

Malignant melanoma is exceedingly rare; only one authentic tumour arising primarily in the meatus has been recorded (Friedmann and Radcliffe, 1954).

Basal cell carcinoma

Basal cell carcinoma arising primarily in the external auditory meatus is rare. It tends to affect the sexes equally and occurs in late middle life. Good results can be obtained by sleeve resection. The prognosis is favourable.

Sebaceous tumours

Sebaceous cell carcinoma is extremely uncommon; fewer than 100 cases affecting any part of the body have been described. These tumours may arise anywhere but their greatest concentration is in the head and neck, mainly on the concha and nose. There are three types:

1. sebaceous adenoma
2. basal cell carcinoma with sebaceous differentiation
3. true sebaceous carcinoma.

One case has been reported affecting the ear (Doble, Snyder and Carpenter, 1981).

Middle ear

Choristoma

A choristoma is a mass of normal tissue at an abnormal site. Seven salivary gland choristomata of the middle ear have been reported - six were in females. The tumours all presented with deafness, usually lifelong, and many patients showed other anomalies of the
middle ear such as absence of the stapes and an abnormal course of the facial nerve. Attempts to remove the tumour were abandoned in most cases because it was attached to the facial nerve, and because other middle ear structures could not be identified.

**Benign adenoma**

Benign adenomata of the middle ear have recently been reported. The tumour is divided equally between the sexes with a maximum age incidence between 40 and 50 years. The main symptom is unilateral progressive deafness, and the principal clinical finding is a conductive hearing loss. The external canal is usually normal, and the tympanic membrane intact in 75% of patients; 25% have a perforation through which the middle ear tumour can often be seen. Preoperative radiology shows a mass in the middle ear or mastoid, but no bone destruction.

Some adenocarcinomata of the middle ear, previously described, may be benign adenomata, which would explain the unusually good prognosis reported for adenocarcinoma in some series.

**Adenocarcinoma**

If glandular tumours of the external ear are rare, such tumours of the middle ear are even rarer. This is interesting since the middle ear is lined by glandular epithelium. Thirteen patients have been reported in the literature with a female predominance and a median age of onset of about 40 years. Deafness, pain and facial paralysis were the presenting symptoms in decreasing order of frequency. Most were treated by mastoidectomy followed by radiotherapy. Prognosis: six out of either patients were alive at 2 years.

**Squamous carcinoma**

Virtually all malignant epithelial tumours of the middle ear are squamous in type. As the tumour grows it causes extensive bony destruction. The petrous pyramid and especially the labyrinth resist invasion longer than other structures. The pathway of least resistance is through the thin roof of the middle ear into the middle cranial fossa. The dura itself provides a strong barrier and the most common pathway of transdural invasion is along the seventh and eighth nerves in the internal auditory canal, and the petrosal nerve. The temporomandibular joint and the parotid gland may be involved relatively early. Involvement of the ninth, tenth, eleventh and twelfth cranial nerves indicates extension into the neck and along the base of the skull. The tumour may also extend along the eustachian tube to the nasopharynx. These clearly preclude resection *en bloc*.

Lymph node metastases occur in 10-15% of cases and about 10% develop a node metastasis later. Distant metastases are rare but have been reported in the liver, brain, lung and bones.

The cause of death in most cases is cachexia due to a combination of intolerable pain, opiates and cranial nerve involvement. Occasionally, invasion of the meninges leads to fatal intracranial complications; erosion of the jugular bulb or carotid artery may cause terminal haemorrhage.
Staging

Neither the International Union Against Cancer (UICC) nor the American Joint Committee (AJC) have developed a stating system for carcinoma of the ear. Based on the criteria used for other sites, the following seems to be a reasonable suggestion:

T1  tumour limited to the site of origin, that is with no facial nerve paralysis and no bone destruction

T2  tumour extending beyond the site of origin indicated by facial paralysis or radiological evidence of bone destruction, but no extension beyond the organ of origin

T3  clinical or radiological evidence of extension to surrounding structures (dura, base of the skull, parotid gland, temporomandibular joint, etc)

Tx  patients with insufficient data for classification, including patients previously seen and treated elsewhere.

This staging system significantly predicts survival in carcinoma of the ear.

Epidemiology

Incidence

Incidence figures for the UK only became available in 1967. The age-adjusted incidence (registration of new cases) rate has remained steady at about 1/million per year for women and 0.8/million per year for men for the 10 years to 1977. The male (M) to female (F) sex ratio was also stable, at about 1:1.2.

Mortality

The age-adjusted mortality rates are similar for each sex but there is a falling trend for men which has resulted in a decrease in the M:F sex ration between 1960 and 1980 from 1.2:1 to about 1:2.  

Mortality from cancer of the middle ear for the 10-year cohorts born around 1881-1921 reveals a marked difference between men and women. Each male cohort has experienced a lower age-specific mortality than the preceding cohort while there has been no apparent change in the age-specific death rates for successive female cohorts. In men, the mortality decreases after 70-75 years of age, whereas the mortality continues to rise for women. The falling trend in mortality for men but not for women may be due to exposure to an occupational carcinogen which was the cause of the relatively high rate in men in the nineteenth century. Also, these men were involved in the 1914-18 war and it has been shown that mustard gas is associated with a higher risk of death from neoplasm of the respiratory tract (including paranasal sinuses) than expected.
Assessment

Clinical

Local assessment is designed to identify the extent of the tumour, and particularly those factors which render the patient incurable.

A history of chronic otitis media suggests a tumour arising in the middle ear, and absence of this history suggests origin in the external canal. Rarely, the history may also indicate aetiological factors such as previous irradiation. Most patients complain of discharge and deafness, but vertigo is rarely seen. Pain, particularly if deep and boring, indicates dural invasion. Clinical examination of the meatus and middle ear and mastoid cavity (if present) demonstrates the tumour.

Facial paralysis, trismus (indicating invasion of the pterygoids or temporomandibular joint), fullness of the parotid gland (indicating spread through the cartilaginous meatus), fullness of the infratemporal fossa, and perichondritis of the auricle are all important physical signs. Cervical nodes should be felt for, especially in the upper deep cervical and pre- and postauricular groups. Lesions of the lower cranial nerves indicate extension of the tumour to the base of the skull.

Laboratory tests

In addition to the usual general laboratory tests, the aural discharge should be tested for glucose to exclude a cerebrospinal fluid leak.

Radiology

Radiological techniques include plain mastoid and temporal bone radiographs, hypocycloidal tomograms in the coronal and sagittal plane, and computerized tomographic (CT) scans. Plain views and tomograms are used to look for erosion of the petromastoid and tympanic bones, whereas CT scans are used to assess soft tissue extension of the tumour upwards and backwards to the cranial cavity and downwards and forwards into the infratemporal fossa.

Most patients give a history of chronic ear infection, so that sclerosis of the mastoid and clouding of the cells are to be expected and are of no diagnostic value. Ragged erosion - often extensive or an unusual site - suggests tumour. An important sign on the lateral mastoid view is erosion of the articular fossa of the temporomandibular joint, and this is present in 30% of patients. Erosion of the bone of the external auditory meatus is best shown by lateral tomograms.

The avascular bone of the labyrinth is relatively unaffected by carcinoma, and erosion of this area with direct invasion of the inner ear is a late radiological feature.

Extension of the tumour anteriorly to penetrate the bony septum separating the middle ear cavity from the carotid artery is of great pathological importance. This is followed by spread around the artery and extension around the eustachian tube towards the postnasal
space. Erosion of the carotid septum and the margins of the bony eustachian tube, and even soft tissue extension of the tumour anteriorly can be demonstrated by tomography and high resolution CT scanning. Enlargement of the retropharyngeal lymph nodes may also be demonstrated by CT scan.

Other routes of spread of the tumour are upwards through the tegmen tympani, backwards through the mastoid air cells and then through the thin plate of bone forming the posterior wall of the pyramid and underlying the lateral sinus. Erosion of these areas may also be demonstrated radiologically. Once the tumour reaches the cranial cavity, the dura is infiltrated and this is rapidly followed by death. It is thus unlikely that any significant extension into the middle or posterior cranial fossa would be shown on a conventional CT brain scan. A carotid angiogram is thus of no value except to demonstrate a blocked lateral sinus. When the carotid artery itself is infiltrated death soon follows. Retrograde jugular venography may be useful to assess the extent of the disease by demonstrating obstruction of the lateral sinus by tumour.

The differential radiological diagnosis of squamous carcinoma of the middle ear includes tuberculous otitis media, malignant otitis externa, and glomus jugulare tumour.

Many clinicians still depend heavily on the submentovertical view. The spine of the sphenoid provides a readily seen and conventional landmark serving the following purposes:

1. Radiological evidence of invasion of the petrosphenoidal region medial to the spine of the sphenoid may indicate a tympanotubal rather than a petromastoid tumour, particularly if symptoms such as oculomotor paralysis, obscure facial pain or trigeminal sensory loss are present.

2. Bone destruction medial to the sphenoidal spine also indicates that cure by any method of treatment is unlikely. The presence of such an extension is an absolute contraindication to radial surgery as the disease is no longer contained within the temporal bone.

3. A submentovertical view occasionally shows destruction of the arch of the atlas due to metastases to the lateral retropharyngeal node with paralysis of the last four cranial nerves.

Clinicians are more interested in eliciting those signs which influence treatment and prognosis. One of the most important of these is dural invasion, and there seems to be no reliable radiological method of assessing this before operation at present.

Treatment

Untreatable patients

Very few papers comment on the patients who are impossible to treat, and those who are perhaps better left alone. Clearly poor general condition may preclude major surgery, and distant metastases contraindicate treatment, but are rare. Lederman (1965) apparently treated all of his 129 patients. Similarly it appears that Lewis (1975) treated every one of his 143
cases. Six of the author's personal series of 62 patients could not be treated: three because of extensive disease, one because of poor general condition and two because of distant metastases.

**Radiotherapy**

This alone has seldom been recommended except at the Christie Hospital in Manchester. The most favoured role for radiotherapy has been in conjunction with surgery, and the usual routine is to carry out a radical mastoidectomy and to follow this by postoperative radiotherapy. The advantages are:

1. reactions are lessened and the patients are made more comfortable
2. higher doses can be given with less risk of complications
3. at lower energy, irradiation is selectively absorbed in bone and cartilage compared to soft tissue, whereas radiation in the higher range is transmitted with relatively little loss.

This treatment produces a crude 5-year survival of about 35%. In most series results for tumours of the external auditory meatus are about 10% better than those for tumours of the middle ear and mastoid. Little attention has been paid to the problem of primary recurrence after failed radiotherapy. This is surprising because the chance of failure of treatment at the primary site has been high in almost all reported cases. In the author's experience, surgery has been of no value for recurrence of a middle ear tumour after radiotherapy, but is occasionally of value for recurrent tumours of the external meatus, particularly of the non-squamous variety.

**The effects of radiation on the ear**

These are well described by Lederman (1965).

**External ear**

1. The was in the external ear canal thickens and dries.
2. The tympanic membrane becomes red and congested, particularly in the region of the handle of the malleus.
3. Oedema may occur particularly antero-inferiorly.
4. Radiation membrane forms along the floor of the external canal, but in the absence of ulceration, widespread membrane formation within the lumen of the canal is rarely seen.
Middle ear

The changes in the middle ear during and after radiation depend on:

(1) The state of the middle ear. The response to treatment is dominated by the changes observed in the neoplasm. Unfortunately, an initial radical mastoidectomy means that the radiotherapist is presented with a cavity in bone devoid of the vasculature essential for the development of the classical responses to radiation. The discharge from the mastoid cavity should diminish with radiotherapy and the bony cavity may become lined with radiation membrane at the end of treatment. In the successful case the cavity should ultimately become quite dry.

(2) The technical method of radiation employed.

Severe reactions and risks of postradiation complications are more likely if large doses are given in a short time by any form of external irradiation or when local contact or implantation methods of treatment are used.

Inner ear

The vestibular apparatus is relatively immune to radiation damage and vestibular symptoms are exceptional. The patient who already suffers from tinnitus, giddiness or sensorineural deafness is uninfluenced for better or worse by irradiation.

Complications of radiotherapy

The external auditory canal

Osteonecrosis of the bony portion of the canal and stenosis are two possible complications. The necrotic bone is usually seen as an exposed area of bone that ultimately forms a scale-like sequestrum. The process is painful and tedious, but healing ultimately occurs. Stenosis of the canal can occur if radiation is given after a sleeve resection or if local radium is inserted into the canal.

The middle ear and mastoid

The possible complications are:

(1) osteonecrosis

(2) damage to the brain, brainstem or eyes.

Osteonecrosis

Osteonecrosis after radiotherapy usually means persistent disease unless the dosage given has been excessive.
**Damage to the brain, brainstem or eyes**

Damage to the eyes should be avoided, but it is impossible to irradiate the petrous temporal bone and much of the middle cranial fossa without irradiating cerebral tissue. Provided that large volumes of tissue are not irradiated to high doses the risk of damage seems to be small. Brain necrosis is more likely to follow radiotherapy in elderly patients whose vasculature is already the seat of arteriosclerotic changes.

The facial or auditory nerves in their extracranial course are never damaged by therapeutic radiation, and any impairment of their function is due either to involvement by cancer or to a postoperative complication if radical surgery has been employed.

**Conductive deafness after radiotherapy**

The loss of hearing following radical mastoidectomy for petromastoid carcinoma is not influenced in any way by radiation. Postradiation conductive deafness may occur in patients who have been successfully irradiated for carcinoma of the external auditory meatus. If the middle ear was normal to start with and the patient is free of recurrence, the possible causes include:

1. thick mucus in the nasopharynx blocking the eustachian opening
2. atresia of the eustachian orifice which is rare and is the result of necrosis of the eustachian cartilage, characterized by severe earache and trismus
3. fibrosis of the fascial space surrounding the levator palati muscle.

**Surgery**

Surgery to the primary tumour may be used in the following ways:

1. as a preliminary measure, usually a radical mastoidectomy, before radiotherapy
2. as a primary form of treatment
3. for salvage after failed radiotherapy.

**Combined surgery and postoperative radiotherapy**

This is the routine used most often and consists of a radical mastoidectomy (extended if necessary), followed as soon as healing is complete by radiotherapy. This was the policy advised by Lederman (1965), a radiotherapist who has almost certainly had the greatest experience of this disease. He only recommended radiotherapy alone for a tumour localized to the external canal not involving the tympanic membrane or the middle ear. Such tumours are rare. For all other tumours he recommended an initial radical mastoidectomy for the following reasons:
1. The temporal bone is one of the densest bones in the skull, and its invasion by tumour is inevitably associated with sepsis which militates against the use of radiotherapy.

2. It is very difficult to assess by clinical or radiological means the full extent of the petromastoid tumour. Preliminary surgical exploration is therefore necessary because this helps to remove necrotic or invaded bone and determine the extent of the tumour.

3. The patient is usually made more comfortable by the operation and loses many of his symptoms.

4. A cavity is provided for drainage and inspection.

The only major authors to demur from this view are the Manchester School who recommend the use of radiotherapy alone.

**Primary surgery**

The first subtotal petrosectomy was reported by Lewis in 1954. He has been the surgeon mainly responsible for developing surgery in this area (Lewis, 1975). The mainstay of surgical treatment is petrosectomy, but as pointed out by Lewis, the operation usually carried out is more than this, and consists of a block resection of the involved portion of the auditory canal, middle ear, mastoid process, petrous bone, temporomandibular joint, parotid gland, base of the zygoma, and usually the auricle and surrounding skin. The highlights of this procedure, mainly taken from Lewis's descriptions are as follows.

(1) Preliminary packing of the lateral sinus. The lateral sinus is exposed by routine cortical mastoidectomy about 10 days before the main procedure and the sinus packed off tightly with BIPP gauze. This induces thrombosis in the venous lake in which the bone lies and drastically reduces bleeding at the main procedure.

(2) Incision and exposure. If the auricle is to be preserved, a U-shaped incision should be made and the auricle reflected upwards. However, the auricle is more usually sacrificed and a large circular incision to include the auricle, and any invaded skin, is then made around the ear.

(3) Temporal craniotomy is made early in the procedure to assess invasion of the dura and the apex of the petrous pyramid. Invasion of a small part of the dura is not an indication of non-resectability, but invasion of the petrous apex is. Cerebrospinal fluid may be removed at this point by a malleable needle to allow the dura and brain to be separated from the underlying bone.

(4) Parotidectomy with tagging of the peripheral branches of the facial nerve if a graft is to be carried out.

(5) Division of the temporomandibular joint and zygomatic arch.
(6) Division of the styloid process to define the position of the carotid artery.

(7) Division of the posterior border of the mastoid process posterior to the lateral sinus.

(8) Division of the floor of the middle ear and the bulb of the jugular vein.

(9) Transection of the petrous pyramid lateral to the internal carotid artery and medial to the arcuate eminence using a Stryker saw or curved chisels.

(10) Cover of the soft tissue defect by grafts, muscle or skin flaps.

(11) Reconstitution of continuity of the facial nerve.

Reconstitution of the facial nerve

The following procedures have been advised.

(1) Facial-hypoglossal anastomosis is recommended by Lewis. He does not quote any results. A lateral tarsorrhaphy should, of course, be performed in addition.

(2) Fascial slings. A cross-face graft using the sural nerve is another other possible technique but does not appear to have been described in this disease.

Soft tissue cover

If the auricle is preserved it is only necessary to apply a skin graft to the mastoid cavity. If the auricle has been resected cover by a skin flap is required. Many authors still favour the use of a local scalp flap based anteriorly or posteriorly and this remains the simplest and most reliable method in the author's experience. A nape of neck flap has been used, but this type of flap has now passed into history. The deltopectoral flap has been advocated by some but it, too, is now obsolete.

With the development of musculocutaneous flaps, particularly the pectoralis major, it is inevitable that they have been used to close this defect. The pectoralis major flap is said to have significant advantages over a scalp flap because it provides a large surface area and adequate bulk to fill defects after removal of the temporal bone. Osteoradionecrosis does not occur because the muscle of the pectoralis major flap provides rich vascularity for healing of an area contaminated with bacteria. Furthermore, the flap can tolerate postoperative radiation, and the muscle provides a seal in the event of a cerebrospinal fluid leak. A further advantage is that reconstruction can be performed in one stage without the need for skin grafts; this causes less pain in the postoperative period and allows earlier discharge from hospital.

Management of lymph node metastases

Lymph node metastases in this disease are unusual. The highest rate quoted is that of 21%, but most authors report a lower incidence. A figure of 10% for tumours of the external meatus would be more usual. All appear to be agreed that there is no place for prophylactic
Complications of surgery

These include:

(1) Bleeding. This can be reduced to relatively negligible amounts by preoperative packing of the lateral sinus.

(2) Infection is common because many of the patients have long-standing chronic otitis media with infection by Gram-negative organisms. Appropriate chemotherapy is therefore required.

(3) Loss of facial nerve function. The facial nerve is divided both in the temporal bone and parotid gland.

(4) Deafness and vertigo. Deafness is complete following this procedure because of the removal of the structures of the inner ear. The patient is also vertiginous for a period of up to 3 weeks until compensation occurs.

(5) Carotid artery thrombosis, may occur due to trauma to the carotid vessels and has, on occasion, led to hemiplegia.

(6) Damage to the lower cranial nerves.

(7) Cerebrospinal fluid leak.

Choice of treatment

The criteria for choosing primary radiotherapy or primary surgery have not been defined; both give a 5-year survival of 30-35%, but the morbidity and mortality from primary surgery are clearly greater. Certainly small tumours without a facial paralysis, bony erosion or lymph node metastases should be treated by radiotherapy. Sadly these patients are uncommon. Patients with disease beyond the temporal bone are almost certainly incurable by either method. It remains uncertain what to do for the patient with disease spreading within, but still confined to, the temporal bone.

The recommended treatment for hidradenomata of the external auditory canal is as follows: local excision for ceruminous adenoma, excision en bloc of the canal and surrounding bone with preservation of the facial nerve for adenoid cystic carcinoma, subtotal petrosectomy and postoperative radiotherapy for ceruminous adenocarcinoma, and excision biopsy for pleomorphic adenoma.
Prognosis

The prognosis for squamous carcinoma of the external meatus is better than that for the middle ear.

Tumours of the external canal may be classified as localized or extensive. The prognosis of the former is of course better. Invasion of the petrous apex, the eustachian tube or the dura indicates incurability. Lymphadenopathy at the time of presentation is a grave prognostic sign. The histological grade of squamous cell carcinoma of the external meatus is not a prognostic indicator.

The prognosis for benign and malignant glandular neoplasms of the external meatus varies with the histological type.

Sophisticated mathematical techniques (generalized linear interactive modelling) are not available to assess the prognostic factors, taking into account both survival and length of survival. These techniques have shown that the staging system suggested above, histological type, the patient's general condition and the presence of lymph node metastases are significant predictors of survival.

Acknowledgements

This chapter is based on a review (Stell, 1984) where a full list of references is available.