Chapter 2: Radiology of the ear, nose and throat

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The principles of technique which apply to imaging in adults apply equally to the demonstration of head and neck lesions in infants and children. Optimum spatial and density resolution with lowest possible level of patient irradiation and freedom from movement artefacts must be achieved. Limitation of radiation dose is particularly important in this age group, and minimal patient movement is hard to obtain. Consequently, many of the more sophisticated modes of imaging such as xeroradiography, conventional tomography and computerized tomography (CT) are used less in the younger age group and most imaging assessments rely on plain films. However, the role of CT continues to increase and, with scan times of a few seconds possible on the latest machines, these examinations can now usually be performed without sedation.

Sedation and anaesthesia in paediatric radiology

A completely immobile patient is necessary for most imaging, especially when there are long radiographic exposure times. This may be difficult to achieve in young patients, especially those who are hyperactive, mentally retarded or disturbed; Hutton (1981) has described a regimen of sedation and anaesthesia for children undergoing tomography and electrocochleography. He recommends ketamine anaesthesia for children under 3 years of age, and conventional inhalation anaesthesia for those above this age.

Upper respiratory tract

Air within the structures of the upper respiratory tract provides a natural contrast medium for the accurate delineation and evaluation of the adjacent soft tissues of the neck. Most of these assessments are made on the lateral neck film obtained during inspiration with the neck partially extended. Additional views can be employed as the need arises, and radiology can often provide information as to the specific cause of airway obstruction and to its site. Well-coned frontal and lateral films are obtained; inspiratory and expiratory films may be needed in each projection to reveal the abnormality fully.

The prevertebral soft tissues and the airway in general are extremely pliable in infants, and with expiration and flexion of the neck a wide variety of distortions and bizarre appearances can result; these are discussed in Chapter 30. High kilovoltage techniques with special filters in the X-ray beam can give detailed radiographs of the upper airway for reduced radiation dose (Joseph et al, 1976). Such techniques can usually obviate the need for xerography. Although xerography can give an enhanced demonstration of the air-soft tissue interface, the radiation dose is higher, and in the opinion of the author little further information is obtained.

Inflammatory disease of the pharynx, larynx and epiglottis are common causes of upper respiratory obstruction and potentially very serious. Radiological examination is rarely necessary to demonstrate these conditions directly but is needed to exclude pulmonary disease. A soft tissue lateral view of the neck may show soft tissue swelling of the larynx, and can reveal the extent of a retropharyngeal abscess.
Conventional tomography is particularly useful in the frontal projection for showing subglottic stenosis and webs. It can be combined with CT to assess congenital abnormalities such as vascular rings or developmental cysts which cause stridor by distortion of the trachea at a lower level.

**Face and sinuses**

The sinuses are present in the infant and can become infected. At birth small antra and ethmoid sinuses can be demonstrated radiographically. The frontal and sphenoid sinuses develop later. The lateral view does not present any problem, but proper positioning is essential to show infant sinuses. The most important plain film sinus view in the adult is the occipitomental with the baseline at 45° to the plane of the film (see Volume 4). In the older child this angle must be decreased to about 28° or the projection will be too steep for proper evaluation of the maxillary antra. In the infant the angle must be even less so that the view is almost a posteroanterior view (Hayden and Swischuk, 1984). Radiographically the findings of sinusitis vary according to age but, as in the adult, loss of the normal radiolucency usually indicates disease. As well as the loss of normal aeration, more specific features such as mucosal thickening and fluid levels may be recognized. Mucocoeles from obstruction of the sinus ostium are rare in children, and are best demonstrated by sectional imaging.

**Choanal atresia**

Choanal atresia is the commonest congenital abnormality of the nose, but cannot be demonstrated by plain film views. Although CT would probably now be the investigation of choice in the first instance, a very satisfactory demonstration of the site of obstruction can be made by instilling an oily contrast medium into the nasal cavity and obtaining films in the lateral and base projections.

**Developmental mass lesions**

Developmental mass lesions affecting the upper respiratory tract include meningoencephaloceles and arachnoid or dermoid cysts. Encephaloceles occur in the midline in the nasofrontal and nasoethmoidal regions. Although they may be demonstrated by conventional imaging techniques, CT scanning is the investigation of choice. This is particularly so for the much rarer lateral protrusions through the base of the skull. They may present as masses in the infratemporal fossa which subsequently expand and bulge into the aerodigestive tract. Dehiscences in the skull base occur with neurofibromatosis and defects in the back of the orbit may result in proptosis.

**Juvenile nasopharyngeal angiofibroma**

Although the pathogenesis of so-called juvenile nasopharyngeal angiofibroma is uncertain and it occurs in an older age group, this vascular tumour also appears to be of developmental origin. Lloyd and Phelps (1986) in a recent study of 30 cases have shown that angiofibroma takes origin at the sphenopalatine foramen. It enlarges the foramen and erodes bone locally: at the base of the medial pterygoid plate, the floor of the sphenoid sinus and the posterior wall of the maxillary antrum. Further extension leads to invasion of the
infratemporal fossa, orbit and middle cranial fossa. Severe bleeding may accompany biopsy and for this reason most surgeons are reluctant to undertake biopsy of a nasopharyngeal mass in an adolescent male patient, and prefer to relay upon clinical and radiological features to decide whether the mass is likely to be an angiofibroma or a non-vascular lesion such as an antrochoanal polyp. Traditionally this involved angiography to show the characteristic vascular blush, supported by the 'antral sign' or indentation of the posterior wall of the maxillary antrum shown on lateral plain films or tomography.

However, the antral sign is not specific for angiofibromata. It can occur with any slow-growing mass in the infratemporal fossa (see Volume 5) and was only positive in 81% of the author's patients with angiofibroma.

A more reliable sign is erosion of the base of the medial pterygoid plate, associated with enlargement of the sphenopalatine foramen which was demonstrated in 100% of 28 patients examined by conventional or computerized tomography. Once the diagnosis is established then the role of the radiologist is to define the limits of the tumour prior to surgery since this may influence the surgical approach. Three-plane magnetic resonance tomography is the method of choice. It best demonstrates the extent of the tumour, it uses non-ionizing radiation, and it will show the vascular nature of the angiofibroma and confirm the diagnosis. Angiography need only be performed if embolization is deemed necessary prior to surgery.

The natural history of angiofibroma, treated or untreated, is highly variable. It has long been known that there is a tendency to involute with age, and these clinical observations have been supported by histological evidence that individual tumours show an increase in fibrous elements with time. However, at least partly because of the difficulty of assessment of the area of origin, and because of the reluctance of clinicians to intervene in the absence of symptoms, there seems to be no firm evidence in the literature that spontaneous involution of untreated lesions occurs (Chandler et al, 1984). Recently, complete regression of a small tumour that persisted after surgical removal of the original mass has been described (Stansbie and Phelps, 1986). Involution of this tumour was fully documented by serial CT scans over a period of 4 years.

Lesions of the jaws

Congenital deformities of the face and jaws usually present to plastic surgeons or to oral surgeons and orthodontists because of bite problems. The two most important of the first arch syndromes are:

(1) Hemifacial microsomia in which there is underdevelopment of one half of the face. In the 20% of cases where the deformity is bilateral there is always dissymmetry between the two sides.

(2) Mandibulofacial dysostosis or the Treacher Collins syndrome, where there is characteristic bilateral and symmetrical hypoplasia of jaw and ear structures. The pathogenesis of both conditions has been discussed by Poswillo (1974).

Underdevelopment of the ascending ramus and condyle of the mandible appears to be the hallmark of hemifacial microsomia, and these abnormalities can be well shown by lateral
and frontal radiographs and especially by orthopantomography. It has long been recognized that this hypoplasia affects not only bony structures but also the soft tissues, and in particular the muscles and the parotid gland. Only recently has the use of CT scanning enabled this soft tissue hypoplasia to be adequately demonstrated. Characteristically there is most underdevelopment of the masseter muscle and the parotid gland, a feature often apparent on clinical examination, although the pterygoid muscles are also commonly affected.

Symmetrical hypoplasia of jaws and muscles is a feature of the Treacher Collins syndrome but this is more uniform and less pronounced than in hemifacial microsomia and the salivary glands are not affected. Antegonal notching of the body of the mandible is a feature of the condition, and crowding of teeth causes orthodontic problems.

**Fibrous dysplasia**

Tumours and tumour-like lesions of the jaws are uncommon in children, and fibrous-osseous abnormalities such as fibrous dysplasia predominate. They cause a painless and slowly developing expansion of the jaws and may have characteristic appearances on plain films or CT, but are more often cystic with non-specific features, although a well corticated margin confirms the benign nature of the lesion. A giant cell reparative granuloma of the upper alveolus is shown in Volume 4.

Rare malignant neoplasms cause extensive ragged destruction of the jaws. Sometimes calcification or even new bone formation within the tumour may be diagnostic, especially in osteogenic sarcoma.

**Bone marrow tumours**

Bone marrow tumours in children not infrequently involve the mandible. Leukaemia, lymphoma and metastatic neuroblastoma can produce mottled destruction of the mandible and destruction or disruption of the teeth. Lytic destructive lesions are a common feature of histiocytosis and the mandible is often involved.

**The petrous temporal bone**

The middle and inner ears are fully developed at birth, but the temporomandibular joint and mastoid process are not. Postnatal changes in the temporal bone consist of growth and pneumatization of the mastoid process and alteration in the shape of the tympanic ring. Prior to full ossification of the petrous pyramid, the dense bone of the labyrinthine capsule can be clearly identified on plain mastoid views, enabling gross developmental abnormalities to be identified without the need for sectional imaging. In the middle ear the ossicles can be shown and, in the neonate, even marrow spaces.

**Congenital ear deformities**

Many congenital abnormalities of the hearing organ do not involve bony structures and therefore cannot be shown by radiological methods. Nevertheless, structural abnormalities of the inner, middle and external ear can be shown in considerable detail by tomographic techniques. Unfortunately, affected children and usually referred between the ages of 2 and
4 years, when the deafness is first confirmed, and sedation or a general anaesthetic is required for the examination. If, after careful consideration, it is felt that the results of the investigation would be unlikely to affect patient management it may be reasonable to defer the examination until the child can cooperate. In the neonatal period a few tomographic sections can usually be obtained for those relevant external deformities or syndromes in which temporal bone abnormalities are a feature. These syndromes with recognized structural abnormalities of the temporal bone are reviewed in Chapter 4 of *Radiology of the ear* (Phelps and Lloyd, 1983).

The purpose of the radiological examination is first to demonstrate any bony abnormality of the inner ear, and particularly of the cochlea. This is complementary to the audiological assessment and ideally electrophysiological studies (either auditory brainstem response or electrocochleography) should be undertaken at the same time. Deformities associated with an actual or potential cerebrospinal fluid fistula may be demonstrated.

Congenital abnormalities of the middle and external ears are shown much more often than deformities of the inner ear, although combined deformities occur in about 20% of cases. The study of the outer ear relates to the prospects for surgical intervention to improve the sound conducting mechanisms and is mandatory before any exploration of congenital atresia. Surgery is now, however, rarely performed for unilateral lesions, but in bilateral atresias the radiological examination is crucial to indicate the best side of exploration.

Sectional imaging with a profound knowledge of normal and abnormal temporal bone anatomy on the part of the observer is required. For bony abnormalities of the inner ears, conventional tomography is just as good as CT, but for the middle ear and external meatus, thin section, high resolution CT with its ability to demonstrate both bone and soft tissue abnormalities is essential before any surgical exploration; particularly for the all-important assessment of the presence, state and size of the middle ear cavity.

**Inner ear deformities**

Congenital malformations of the bony labyrinth, internal auditory meatus and vestibular aqueduct, which vary widely in severity from minor anomalies with normal cochlear function to severe deformities which preclude any level of hearing whatever, may be suggested by audiological assessment. Traditionally two eponyms are enshrined in accounts of congenital deafness and so need to be defined:

1. Michel defect (Michel, 1863) - complete lack of development of any inner ear structures.

2. Mondini defect (Mondini, 1791) - a cochlea with one and a half turns and the apical coil replaced by a distal sac. Although the subject of Mondini's dissection had been completely deaf, the normal basal turn of the true Mondini defect means that some hearing is possible. Mondini's case also had very dilated vestibular aqueducts (Phelps, 1986).

Line drawings of some examples of labyrinthine deformities are shown. A primitive sac with one or more appendages is commoner than a Michel deformity.
The semicircular canals may be missing or dilated in varying degree, but the commonest inner ear anomaly, namely a solitary dilated dysplastic lateral semicircular canal is often associated with normal cochlear function. Dilatation of the vestibular aqueduct often accompanies minor abnormalities of the bony cochlea and vestibule and congenital hearing loss (Valvassori, 1983). The deafness may be fluctuant and/or progressive giving rise to speculation that endolymphatic hydrops is also a feature.

Anomalies of the internal auditory meatus include the bulbous type which is usually of no significance; unusual direction which is the result of skull base aberrations; and very narrow or double internal auditory meatus which usually indicate severe or total deafness (Phelps and Lloyd, 1983).

**Inner ear lesions associated with cerebrospinal fluid fistula**

Congenital cerebrospinal fluid fistula into the middle ear cavity is a rare but potentially fatal condition which is frequently misdiagnosed. When the fistula occurs spontaneously it usually presents in the first 5 or 10 years of life as:

1. Cerebrospinal fluid rhinorrhoea if the eardrum is intact. Cerebrospinal fluid passes down the eustachian tube causing a nasal discharge.

2. Cerebrospinal fluid otorrhoea if there is a perforation in the eardrum, or if myringotomy has been performed for presumed serous otitis media.

3. Attacks of meningitis which are usually recurrent. At times meningitis is the sole presenting manifestation of a cerebrospinal fluid fistula.

Deafness is usually severe or complete, but it is difficult to diagnose and assess, especially in a young child. It is frequently unrecognized if unilateral. The conductive and sensorineural components of the deafness are also hard to define.

Spontaneous cerebrospinal fluid fistulae from the subarachnoid space into the middle ear cavity may be classified as perilabyrinthine or translabyrinthine. The very rare perilabyrinthine group, through bony defects close to but not involving the labyrinth, usually have normal hearing initially. The commoner translabyrinthine group is nearly always associated with anacusis, severe labyrinthine dysplasia and a route via the internal auditory meatus. The labyrinthine deformity is more severe than the type classically described by Mondini, and evidence of a dilated cochlear aqueduct in these cases is also unconvincing.

The perilabyrinthine and translabyrinthine routes are discussed in a recent paper by Phelps (1986). The most important route is via an abnormally shaped internal auditory meatus that usually tapers at its lateral end. The cochlea is an amorphous sac which lacks a modiolus or central bony spiral. The cochlear sac may be bigger or smaller than a normal cochlea. No proper basal turn can be recognized as in a true Mondini deformity, and there is a wide communication between the cochlear sac and the vestibule which is itself abnormal and enlarged, especially in the horizontal plane. The semicircular canals may be dilated to a varying degree, especially the lateral.
The labyrinthine malformation is often accompanied by a defective stapes - usually a hole in the footplate - and the exit route of cerebrospinal fluid into the middle ear is via the oval or, less commonly, the round window. It should be stressed that the fistula is usually spontaneous or the result of a minor head injury.

Congenital fixation of the stapes footplate is likely to be associated with a profuse perilymph or cerebrospinal fluid leak following stapedectomy. The surgical results of stapedectomy for congenital fixation are not very satisfactory, but there is little radiological evidence of structural abnormalities of the labyrinth in these 'gushers'.

The management of cerebrospinal fluid fistulae into the middle ear depends on a high degree of clinical suspicion. Perilabyrinthine fistulae are extremely rare and usually associated with normal hearing. Bone defects around the labyrinth may be shown by sophisticated bone imaging, but tracer cerebrospinal fluid contrast studies may be necessary to confirm the aural route. The commoner translabyrinthine type is almost always associated with labyrinthine dysplasia. Sensorineural deafness or two unexplained attacks of meningitis make a polytomographic or CT study of the temporal bones mandatory.

**Middle ear deformities**

Radiology of congenital deformities of the middle and external ear relates almost exclusively to the prospects of improvement of conductive deafness by surgical intervention. The size and shape of the middle ear cavity is the most important assessment to be made, especially where there is atresia of the external auditory meatus.

In the majority of unilateral atresias with associated deformity of the pinna but no other congenital abnormality, there is a normally formed mastoid with good pneumatization and the middle ear cavity is of relatively normal shape. Even in the most severe deformities there is rarely complete absence of the middle ear and usually at least a slit-like hypotympanum can be shown lateral to the basal turn of the cochlea. The middle ear cavity may be reduced in size by encroachment of the atretic plate laterally, by a high jugular bulb inferiorly or by descent of the tegmen superiorly. In craniofacial microsomia and mandibulofacial dysostosis, the attic and antrum are typically absent or slit-like, being replaced in varying degrees by solid bone or by descent of the tegmen.

If the middle ear cavity is air containing, its shape and contents are relatively easy to assess. Frequently, however, the middle ear in congenital abnormalities contains undifferentiated mesenchyme, a thick glue-like substance which is radiologically indistinguishable from soft tissue or retained mucus. This bony septa may divide the middle ear cavity into two or more compartments.

**Facial nerve**

The next important structure from a surgical point of view is the facial nerve. The nerve is very rarely absent, although it might be hypoplastic. The main problem is aberration in the course of the nerve.
In early embryonic life, the developing seventh cranial nerve lies anterior to the otocyst, so if development is arrested at this stage, a tract for the facial nerve is found anterior to a primitive otic sac. If development is arrested at a later stage, after the cochlea has formed to some extent, then the first part of the facial nerve is found in its usual situation above and lateral to the cochlea. The facial nerve is, therefore, relatively unaffected by developmental abnormalities of the labyrinth, and aberrations of the first part of the facial nerve canal are most unusual.

The course of the second and third parts is, however, dependent on normal development of the branchial arches, the facial nerve being the nerve of the second arch. During its development and migration, the facial nerve curves behind the branchial cartilage to reach the anterior aspect of the same cartilage. At the same time, part of the cartilage adheres to the otic capsule to form the fallopian canal. If, during development, the external pharyngeal groove of the first branchial arch is active and atresia is due only to maldevelopment of the tympanic ring, then the second and third parts of the facial canal follow a relatively normal course. In major atresias, when the external pharyngeal groove is not active, then development of malformations is much worse. The temporomandibular joint may abut directly onto the mastoid process.

The greater the deformity the more marked is the tendency for the facial nerve to follow a more direct route out into the soft tissues of the face. Exposed facial nerves in the middle ear cavity are the most common abnormalities recorded at surgery for congenital malformations. Usually the fallopian canal is dehiscent but the descending segment may also be exposed, and overhang of the facial ridge with absence of the second genu is a usual finding in the Treacher Collins syndrome making access to the oval window difficult for the surgeon. A short vertical segment of the facial canal and high stylomastoid foramen mean that the nerve turns forwards into the cheek in a high position.

In the preoperative radiological assessment the descending facial canal and its relationship to other structures must be demonstrated, preferably in both coronal and lateral sections. Axial CT sections will show the descending canal in cross-section and identification is less certain. Grossly displaced nerves that cross the middle ear cavity are more difficult to identify even by CT, but two useful signs of aberrant pathways through the middle ear cavity are:

1. An exit foramen through the floor of the middle ear cavity or lateral atretic plate may be identified.

2. Absence, at the back of the middle ear cavity, of the pyramidal eminence which normally contains the stapedius muscle and tendon (see Volume 3, Chapter 2).

The pyramid was identified in only two of the cases noted at operation to have anomalies of the facial canal in the middle ear and, in these, the dehiscence was in the second part. Absence of the pyramid is, therefore, good presumptive evidence of an exposed facial nerve. Bifurcation of the descending portion is far commoner with congenital malformations than in normal patients.
Ossicles

A normal ossicular chain is rarely found where there is atresia of the external ear, but complete absence of the ossicles is also unusual. In most cases at least some vestige of the ossicular chain is evident. The ossicles are often thicker and heavier than normal or, less frequently, thin and spidery. They may be fixed to the walls of the middle ear cavity by bosses of bone but the more usual deformity discovered at surgery is a fusion of the bodies of malleus and incus. The ankylosis varies in degree and may be bony or fibrous. The radiological recognition of this ossicular union is difficult but is, in any case, not of great practical importance and an irregular lump of bone in the middle ear cavity usually represents an ossicular mass.

Because of the partial or complete replacement of the tympanic membrane by a bony plate, the handle of the malleus is not surprisingly that part of the chain which is most often abnormal and most easily recognized on the tomograms. If the handle is absent the 'molar tooth' appearance of the ossicles will no longer be evident in the lateral projection and a triangular appearance of the ossicular mass will be seen. Often, the handle of the malleus is bent towards the atretic plate to which it may be fixed and this gives the typical L-shaped appearance to the ossicular mass. A slit-like attic so typical of Treacher-Collins syndrome or an overhanging facial ridge may obstruct the free movement of the ossicular chain.

External auditory meatus

In congenital deformities of the external ear, the external auditory meatus may be narrow, short, completely or partially atretic or it may run in an abnormal direction. It often slopes up towards the middle ear and in such cases it may be curved in two planes, becoming more horizontal at its medial end. The obstruction in atresia may be due to soft tissue or bone but usually both are involved. The tympanic bone may be hyperplastic (rarely), deformed or absent.

The so-called atretic plate may therefore be composed partly of a deformed tympanic bone and partly of downwards and forward extension of squamous temporal and mastoid bones, in which case it may be pneumatized.

A diagrammatic representation of some of the congenital structural abnormalities of the middle and external ears as shown by coronal section imaging is given.

 Syndromes

It is not intended to discuss the radiological features of syndromic ear deformities except for the two commonest and most important.

Hemifacial microsomia

The ear lesions are usually bizarre and severe. The pinna is often represented by a small tag. Meatal atresia and middle ear abnormalities are almost constant findings and there may be gross descent of the tegmen to, or even below, the level of the lateral semicircular canal. Occasionally, some degree of hyperplasia of external ear structures, particularly the
tympanic bone, occurs but the mastoid is hypoplastic and unpneumatized. The middle ear cavity is usually small, being encroached upon by the low tegmen and thick atretic plate. The ossicles in such cases are absent or hypoplastic and malformed. Three of the author's patients had an ossicular mass displaced laterally, far from the oval window. This anomaly is only seen in cases of facial microsomnia. The condition is not exclusively unilateral and often involves the bones of the skull base. Though bilateral, there is always considerable dissymmetry between the two sides. This dissymmetry distinguishes the syndrome from Treacher Collins syndrome, with which it has often been confused in the past. There is no hereditary factor in craniofacial microsomnia. It is the most common of the otocraniofacial syndromes (Phelps, Lloyd and Poswillo, 1983).

**Treascher Collins syndrome**

The middle ear abnormalities in Treacher Collins syndrome are symmetrical and characteristic, although they may vary in severity (Phelps, Poswillo and Lloyd, 1981). The mastoid is unpneumatized and the attic and antrum are often reduced to slit-like proportions. Atresia of the external auditory meatus is a less constant feature and in 50% of patients the meatus may be patent, although it tends to be curved, running upwards in its lateral part. Ossicular abnormalities are common and, in nearly all the operated ears in the author's series, the facial nerve followed a more direct path with opening out of the bends. It usually appeared at surgery as an overhanging facial ridge.

**Bone dysplasias**

Deafness is a common childhood feature of rare congenital generalized bony dysplasias. Only a brief account of the radiological features of osteogenesis imperfecta and of the dysplasias with increased bone density is given here. For more extensive descriptions other works need to be consulted (Booth, 1982; Phelps and Lloyd, 1983).

Deafness in osteogenesis imperfecta tarda may be conductive, sensorineural or mixed. The radiological appearances consist of demineralization in the labyrinthine capsule indistinguishable from otospongiosis but, in contrast to otospongiosis which only affects the capsule, dehiscent ossification occurs in other sites in the petrous pyramid.

The osteopetroses are a group of uncommon genetic disorders that are characterized by increased skeletal density and abnormalities of bone modelling. Common to all of these disorders, is a proclivity for involvement of the calvarium and skull base. An associated constellation of neurological symptoms may result, presumably secondary to bony encroachement on the cranial foraamina. Sectional imaging of the petrous temporal bones shows generalized sclerosis and narrowing of the internal auditory meatus. Encroachment by bosses of bone in the attic may also be revealed.

**Otitis media**

Radiology has little part to play in conditions such as otitis media, which is essentially a clinical diagnosis. Loss of aeration of the middle ear cleft and mastoid may suggest infection, but cell wall breakdown (coalescent mastoiditis) and abscess formation in mastoiditis are hard to demonstrate on plain films, and are better shown by CT.
Cholesteatoma in children

Cholesteatoma in childhood is less common than in adults but is a more aggressive and dangerous disease and shows some special features. Often, large cholesteatomata are associated with small perforations of the eardrum and well pneumatized mastoids (Jahnke, 1982). The cholesteatomata in the author's patients appeared to be primary attic lesions and are considered to be 'acquired' even when the eardrum was intact. There was usually no bone erosion seen upon radiological examination.

While not wishing to discuss the aetiology of cholesteatoma, it does seem that a high proportion of childhood cholesteatomata affecting the middle ear have a congenital origin with an intact, unscarred eardrum and no bone erosion. Characteristically they are localized in the mesotympanum and later extend to the attic. Any discussion of congenital childhood cholesteatoma is complicated by two phenomena which are difficult to explain:

1) Cholesteatoma of the base of the skull not affecting the middle ear cavity, and therefore undoubtedly of congenital origin, is predominantly a disease of later life. These lesions from presumed squamous cell rests are therefore considered in Volume 3, Chapter 2.

2) The external auditory meatus develops by recannulation of a solid plug of ectodermal cells of the first branchial arch. This process begins at the medial end, with the membrane separating the primitive meatus from the tubotympanic recess developing into the eardrum. Failure of recannulation will result in congenital atresia of the external auditory meatus, potentially with the epidermis trapped medial to the atretic plate. This situation would seem to have all the potential for development of a cholesteatoma, especially if, as is often found at operation, there is a vestigial eardrum present.

Congenital atresia of the external auditory meatus is not rare but, surprisingly, cholesteatoma beyond the atresia is most unusual. Phelps, Lloyd and Sheldon (1977) found only four cases of cholesteatoma or retained squamous desquamation in 270 congenital deformities of the middle and external ear. Of these four cases, two were due to stenosis of the external auditory meatus and two were true soft-tissue atresias. Unfortunately, because the cholesteatoma is radiologically indistinguishable from soft tissue, these could not have been diagnosed even in retrospect. Other authorities have had similar experience with cholesteatoma, in association with congenital ear malformations. It is probable, that in such cases lack of a stimulus, such as infection, means that any squamous cell rests remain dormant.

Neoplasms

Occasionally tumours such as rhabdomyosarcoma or tumour-like conditions such as Hand-Schüller-Christian disease (histiocytosis X) may affect the temporal bone or mastoid. Massive destruction of bone is the usual feature.