Pathological lesions of the external auditory meatus: a review

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Summary
A closer study of the pathology of the external auditory meatus shows that there are various non-neoplastic and neoplastic diseases of local origin which are of great clinico-pathological importance. These include Kimura's disease and angiolymphoid hyperplasia with eosinophilia; progressive necrotizing (or malignant) external otitis. Osteoma and ceruminoma are typical neoplasms of the external auditory meatus which is also the site of polypoid granulomatous or neoplastic structures arising from the middle ear cleft. These so-called 'aural polyps' often dismissed as of little or no diagnostic importance, may contain significant clues to a serious underlying infection or to a malignant neoplasm. Profuse bleeding may ensue on removal of such polyps.

The histopathological features and their significance are described and discussed.

Introduction
The pathology of the ear has attracted scant attention from pathologists who, with a few exceptions, have fought shy of the apparently barren regions of the temporal bone1. There is more to the 'ear' than meets the eye and even the comparatively limited area of the external ear, consisting of the auricle and the external auditory meatus may be the site of a variety of non-neoplastic or tumour-like lesions and true neoplasms.

Some of the varied lesions affecting the external ear may be of local origin, eg malignant external otitis, Kimura's disease. More often the external auditory meatus forms a portal of exit for diseases of the middle ear presenting as 'aural polyps'. Aural polyps have often been dismissed as of little account. Only when all aural specimens are subject to routine histopathological examination is it possible to appreciate that the granulation tissue forming the polyp may reveal some significant clues to complications of chronic otitis media: cholesterol granuloma, tympanosclerosis, epidermoid cholesteatoma, 'glue ear'. Moreover like the tip of an iceberg, it may disguise some underlying processes and/or unfamiliar disease: specific granulomas, Wegener's granuloma, neoplasms (which may bleed profusely on removal of the aural polyp) (Figure 1).

Chondrodermatitis nodularis helicis
Nodular chondrodermatitis of the auricle is an inflammatory lesion of traumatic origin. There are foci of non-specific inflammatory granulation tissue in the subcutaneous tissue forming small nodules that give the condition its name.

Relapsing poly chondritis
This is a rare disorder of cartilage characterized by recurrent prominent inflammation of the cartilaginous tissue of different organs, primarily auricle nasal septum and larynx. Its aetiology is obscure, but certain laboratory observations suggest an immune pathogenesis. The diagnosis is based on the clinical features and the histological changes of chondritis.

Angiolymphoid hyperplasia with Eosinophilia (ALHE) and Kimura's disease
Several hundred cases have been reported in Chinese and Japanese patients and in the West since Kimura et al. described the lesion in Japan2. Wells and Whimster3 were the first to describe nine cases of persistent subcutaneous nodules of the head and neck as 'Subcutaneous angiolymphoid hyperplasia with eosinophilia'. These benign lesions are characterized by the development of single or multiple subcutaneous nodules on the head and neck. Both lesions show male preponderance. ALHE lesions form multiple small dermal papules or nodules of shorter duration affecting older patients. Kimura's disease affects younger persons as a deep-seated large soft-tissue mass of longer duration. The regional lymph nodes are enlarged in contrast to ALHE4.

Histopathology
The nodules occur in the dermis or subcutaneous tissue and are characterized by intensive angioblastic lymphoproliferative activity, forming branching capillaries lined by hyperplastic endothelial cells surrounded by lymphocytes.

Frequently the terms ALHE and Kimura's disease have been used synonymously. However, it is believed that there are significant clinical and pathological features which distinguish Kimura's disease from...
angiolympoid hyperplasia with eosinophilia. The pathogenesis of ALHE is unknown.

**Progressive or necrotizing external otitis**

Progressive or necrotizing external otitis is a severe aggressive infection of the external auditory meatus which affects mainly diabetic patients and is caused by *Pseudomonas pyocyanea*.

The disease affects mainly the elderly, 55 years of age or older. Factors that predispose to the development of the disease are, in addition to diabetes mellitus, old age, minor trauma to the external auditory meatus, blood diseases. It has been reported also in a non-diabetic patient.

The typical sites of entry of *Pseudomonas pyocyanea* into the bone are: (1) at the junction of the cartilaginous and osseous portion at the floor of the external auditory meatus and (2) in the angle of the roof of the meatus and the tympanic annulus. The infection spreads from here in all directions through the temporal bone.

The disease causes serious complications such as extension of the infection to cartilage, bone, nerve and soft tissue resulting in osteomyelitis of the skull, multiple cranial nerve palsies and death. The most common complication and presenting sign is facial nerve palsy.

Light microscopy shows necrosis of the bone of the floor of the meatus covered by acute or chronic inflammatory granulation tissue. Once osteomyelitis of the temporal bone has developed, the disease may spread into the base of the skull. Progressive external otitis had a mortality rate of 25-67% before the advent of efficient antibiotics. When treated properly in its early stages, good results can be obtained so that early diagnosis is important in order to achieve complete cure.

**Neoplasms of the external auditory meatus**

Two neoplasms of the external auditory meatus have attracted some considerable interest and will be discussed here.

**Osteoma**

The commonest of the osseous tumours of the temporal bone that come to the attention of the otologist are undoubtedly the osteomas (or exostoses) of the external acoustic meatus. Most of these growths arise from the tympanic ring and so adjoin the tympanic membrane; they present as single or multiple often bilateral nodules. Microscopically they range in structure from dense sclerotic bone (ivory or eburnated osteoma) to soft spongy bone (cancellous osteoma).

In 1938, Van Gilse reported an increased incidence of ear canal exostosis in active cold-water swimmers. The enhanced popularity of surfing has produced a marked increase in the incidence of ear canal exostosis (swimmer's ears). New bone formation on the inner surface of the tympanic bulla following experimental irritation of the external auditory meatus with cold water was described.

**Tumours of the ceruminous glands**

The actual rate of incidence of these comparatively rare neoplasms is almost certainly higher than the number of published cases may suggest. The tumours arise from the apocrine ceruminous glands and grow slowly so that they may remain unnoticed for years.

Their characteristic epithelial structure consists of an internal layer of columnar cells and a basal myo-epithelial layer resting on a well-defined basal lamina (Figure 2). The cells produce granular secretions and may have apocrine snouts. Their microscopical appearances are similar to those of sweat gland tumours elsewhere and they show the same range of structural variation. Wettl et al. (1972) suggest a useful classification of the predominant morphological types of ceruminoma. These are (1) ceruminous adenoma; (2) ceruminous adenocarcinoma; (3) pleomorphic adenoma (mixed tumour) and adenoid-cystic carcinomas; and (4) cribriform adenocarcinomas (Figure 2).

The cribriform tumours, which are also known as adenoid cystic carcinomas, adenocystic carcinomas and cylindromas, are the least rare. The behaviour of the neoplasms arising from the ceruminous glands of the external auditory meatus is unpredictable from their microscopical features and the present author has classified them as 'intermediate neoplasms'. Invasiveness appears to be the only definite evidence of malignancy. Opinion, however, has hardened in favour of regarding all such tumours as potentially malignant irrespective of whether or not invasive growth has been identified in the biopsy specimen.

The prognosis of all varieties of these tumours is variable. The acceptability of the name ceruminoma, which is commonly given to these tumours has been disputed.

**Tyrosine crystals in ceruminoma**

Tyrosine crystals in salivary gland tumours have been occasionally reported and were described in a ceruminous gland tumour of the external auditory meatus.

**Malignant mesenchymatous neoplasms**

Malignant mesenchymatous neoplasms are rare. Fibrosarcoma of the auricle has been reported. The auricle may be affected by Kaposi's angiosarcoma which is among the most commonly observed oto-logical manifestations of AIDS. Chondrosarcoma and osteosarcoma may arise in the bony wall of the meatus or elsewhere in the aural parts of the temporal bone.

Rhabdomyosarcoma may arise in the auricle; more often it presents in the external auditory meatus as an aural polyp occurring almost exclusively in children (Figure 3).
Pathology of ‘aural polyps’
It is not surprising that any lesion of the external auditory meatus or of the middle ear may present in the shape of a polypoid structure, obstructing the lumen and/or protruding through the tympanic membrane. Any of the neoplasms mentioned above may, in fact, present as a ‘aural polyp’ and attempts at removing it can be accompanied by profuse bleeding. The actual site of origin of the neoplasms of squamous cell carcinomas presenting in this fashion, may be difficult to determine.

However, more aural polyps are of inflammatory origin, composed of non-specific inflammatory granulation tissue. These simple aural polyps can offer microscopical clues to some underlying complications of chronic otitis media. Others may reveal clinically unsuspected diseases such as specific granulomas, Wegener’s granulomatosis and some exotic lesions.

Cholesterol granuloma
Cholesterol granuloma forms part of the inflammatory process of otitis media and can more accurately be identified or confirmed only by the histopathological examination of the specimen removed at operation.

The characteristic features are those of chronic inflammatory granulation tissue containing large numbers of rhomboid clefts of cholesterol crystals dissolved during processing, surrounded by foreign body giant cells or occasionally, xanthomatous cells. There is usually some clear evidence of recent and/or old haemorrhage with haemosiderin pigment (Figure 4).

Epidermoid cholesteatoma
The granulation tissue of the aural polyp contains accumulations of keratin surrounded by foreign-body giant cells (Figure 5). This feature may be linked with an underlying epidermoid cholesteatoma of the middle ear. A recent retrospective study of 100 aural polyps from 96 patients has concluded that any polyp that is composed of granulation tissue containing keratin in flakes or masses has a 70-80% possibility of being associated with an underlying cholesteatoma.

‘Glue ear’
Aural polyps covered by secretory epithelium and containing large gland-like structures filled with PAS-positive mucoid secretion often indicate a similar reaction of the mucosa of the middle ear, leading to an accumulation of mucin in the tympanic cavity.

The goblet cell population of the middle ear mucosa in chronic otitis media contributes greatly to the amount of mucoid secretion and forms the basic substrate of the glue ear syndrome or catarrhal otitis media.

It may be of interest to recall that the role of mucous membranes was first described by Schneider (1614–1618), who had disproved the theory that mucus originated in the pituitary and developed the ‘catarrhal’ theory based on the qualities of mucus to flow and to be accumulated in cavities. Since mucoid secretion is an important element of the glue ear syndrome, the term ‘catarrhal’ has an historical pedigree.

Tuberculosis of the middle ear
Mycobacterial species once played a prominent part in the causation of chronic otitis media. In 1915 Mycobacterium tuberculosis was the cause of 50% of cases in children under one year of age. With the advent of specific antituberculosis therapy, early diagnosis has become more important than ever; and despite its comparative rarity, this condition should
always be considered in the differential diagnosis of otitis. Many of the cases which present primarily with aural symptoms do not display specific clinical features; instead, the symptoms are often those of any non-specific, acute or chronic otitis media.

Sporadic cases of tuberculous middle-ear infection continue to occur presenting as aural polyps formed entirely by tuberculous granulation tissue. However, the diagnosis depends not only on the classical histological picture but, more importantly, on the isolation of the causal organism by culture and guinea pig inoculation.

Attention has been drawn to the fact that a variety of conditions may give rise to a 'tuberculous granuloma'; the commonest being sarcoidosis, Wegener's granuloma, fungal or nocardial infection which may present as an aural polyp. It must be emphasized that the finding of tuberculoid lesions calls for extensive investigation.

Wegener's disease
The middle ear is a rare site of the initial lesions in Wegener's disease, a condition likely to be missed. The middle ear becomes obstructed by giant-cell granulomatous tissue which may protrude into the external auditory meatus. Vasculitis may be present.

In conclusion it cannot be emphasized too strongly that all aural polyps should be submitted for histopathological examination. Selectivity has been advocated as with other 'trivial' specimens but this might not only delay the correct diagnosis but affect the course of treatment.

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