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Pathology in Focus

Schwannoma of the chorda tympani nerve

S. T. Browning, M.Phil., F.R.C.S. (orl-hns), J. J. Phillipps, F.R.C.S., N. Williams, F.R.C.Path.*

Abstract
We report a case of schwannoma of the chorda tympani. This is a very rare benign tumour and only five other cases have been found in the literature. This is the first case to mimic a cholesteatoma presenting as a pearly tumour in the postero-superior segment of the drum with aural discharge and conductive deafness. Diagnosis is usually by biopsy and treatment is surgical with preservation of facial and auditory function. A summary of the other presentations of this tumour together with a review of the histopathology of the disease is presented.

Key words: Schwannoma; Chorda tympani nerve

Case report

A 51-year-old Caucasian woman presented with a two-month history of right-sided deafness and one week of scanty aural discharge. Other ear symptoms were absent. The clinical appearance was that of a keratin pearl arising in the postero-superior portion of the deep ear canal and almost filling the ear canal. Audiometry was within normal limits bilaterally. An examination under anaesthetic allowed a biopsy to be taken. During the procedure clear yellow fluid escaped from the cyst suggesting the presence of a cholesterol granuloma. Histological examination of the biopsy showed keratinizing squamous epithelium with an underlying spindle-cell tumour (Figure 1). There were Verocay bodies (areas of nuclear palisading separated by eosinophilic material) present. Immunohistochemical studies showed positive staining of these spindle cells with antibodies directed against S-100. This is an acidic protein found in glial cells (and also seen in melanocytes, chondrocytes, adipocytes and myoepithelial cells). The appearances were of a benign schwannoma.

On review of the patient two weeks later there was no evidence of the canal polyp. However there was a suggestion of a mass behind the tympanic membrane in the postero-superior quadrant. Computerized tomography (CT) and MRI scanning of the ear did not show any abnormality. A tympanotomy was subsequently performed and revealed a firm pale tumour arising from the posterior half of the chorda tympani nerve between the posterior canaliculus and the incus. This was successfully removed without disruption to the ossicular chain or facial nerve weakness. The patient has made an uneventful recovery.

Discussion

Only four reports detailing five cases of this rare tumour have been found in the literature since 1966.[1-4] Of the cases so far reported all have had a conductive deafness as the main presenting complaint and this has been associated with tinnitus in three cases. There has been no other case presenting with discharge. Typically, vertigo and otalgia are absent. With the exception of one case,[2] all patients have been between 18 and 25 years of age.

The clinical appearance of the tumour varies but in general a firm swelling arising from the postero-superior quadrant of the drum or posterior canal wall is the main finding. This is generally skin covered but one has had an unusually vascular appearance.[1]

Diagnosis of the tumour before biopsy is very difficult[1] and, despite the risks of biopsying neural tissue arising in the ear, this has been the principal method of diagnosis in all cases reported. CT scanning has proven useful in the planning of surgery for tumours that are large but in the current case scanning was normal and the tumour small.

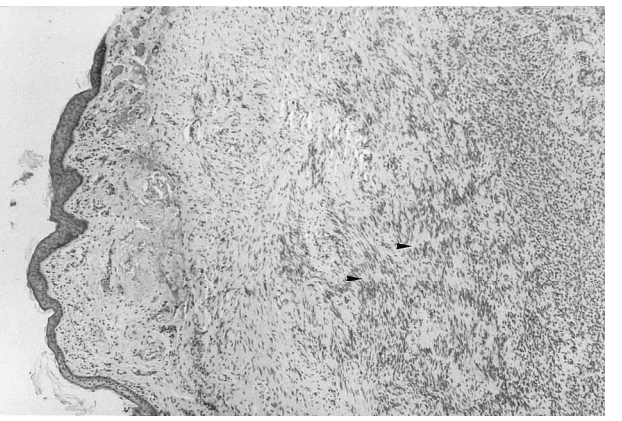

Fig. 1

Histological section of the tumour. There is keratinizing squamous epithelium on the left. An Antoni B area is seen adjacent to the epithelium and a cellular Antoni A area is present to the right. Verocay bodies are indicated by arrows (H&E; ×100).

From the Departments of Otolaryngology – Head and Neck Surgery, and Histopathology*, Singleton Hospital, Swansea, UK.
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Benign schwannomas are common tumours that occur most commonly in middle-aged patients. There is a female preponderance. They are slow growing tumours most common in the head and neck and on the flexor aspects of the limbs. They primarily affect sensory nerves. An association with Von Recklinghausen's disease is rare unless the tumours are multiple. Malignant transformation is very rare. Histologically, schwannomas are spindle-cell tumours characterized by alternating Antoni A and B areas with positive S-100 immunocytochemical staining. The Antoni A areas are cellular with closely packed spindle cells and may contain Verocay bodies. Antoni B areas are less cellular with spindle cells arranged haphazardly in a loose matrix. There may be areas of cystic degeneration within the tumour and it is suggested that it was this that gave rise to the aural discharge and accounted for the yellow fluid expressed from the mass during biopsy.

Conclusion

Schwannoma of the chorda tympani is a rare benign tumour that is difficult to diagnose without biopsy. It may mimic cholesteatoma. Treatment is surgical and recurrence rate.

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Address for correspondence:
S. T. Browning,
Department of Otolaryngology – Head and Neck Surgery,
Singleton Hospital,
Swansea.

Fax: 01792 208647
E-mail: roro@dircon.co.uk