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Salivary gland choristoma of the middle ear

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Abstract

A salivary gland choristoma is an extremely uncommon tumour in the middle ear space. It appears to be a developmental abnormality and may be associated with abnormalities of adjacent structures.

It usually presents with unilateral conductive deafness which may be long-standing and the tumour often pursues a benign, slow growing course. It is usually possible to excise it, but problems may arise as there may be variable associated anatomical abnormalities of the middle ear.

We present the nineteenth recorded case, review the literature and discuss the management of this condition.

Introduction

Choristomas are non-neoplastic proliferations of histologically normal tissue occurring at abnormal sites. They differ from hamartomas in that hamartomas are non-neoplastic proliferations of histologically normal tissue occurring at sites where that tissue is normally found. Salivary gland choristomas have been recorded in the middle ear in both sexes and over a wide age range.

We present the nineteenth recorded case with a review of the literature and discuss the aetiology and management.

Case report

A 10-year-old male was seen in the out patient department with a five year history of decreased hearing in the right ear which lead to the referral by the patients general practitioner. There was no history of ear infections, otalgia, tinnitus or vertigo. The only relevant past medical history is that he had an adenoidectomy in 1985 for persistent nasal obstruction. At that time his tympanic membranes were recorded as normal.

On examination the left ear was normal. On the right the pinna and external auditory meatus was normal. The tympanic membrane was erythematous and it was possible to make out a whitish mass behind the postero-superior segment. Tuning fork tests were consistent with a right conductive deafness. The rest of the examination was unremarkable.

Pure tone audiometry on the left was normal. On the right there was a 50–60 dB conductive loss with normal bone conduction thresholds.

The child was admitted in September 1990 for exploratory tympanotomy. This revealed a pink soft tissue mass covering the promontory extending into the facial recess, covering the incudostapedial joint, and extending into the anterior mesotympanum, medial to the handle of the malleus. The mass was separate from the facial nerve. The long process of the incus was eroded and the round window was completely obscured.

A biopsy was taken as a firm diagnosis was required before proceding to definitive surgery. The histology showed a small glandular structure with an associated duct (Fig. 1). Serous and mucinous secretory elements were present, the pattern resembling that of a minor salivary gland. The duct had a two-layered epithelial structure with a columnar lining. In addition there were scattered similar acini, both serous and mucinous, (Fig. 2), surrounded by collagenous connective tissue within which were striated muscle fibres, blood vessels and nerves. The histological picture was that of disorganized salivary gland-type structures with a connective tissue stroma and was diagnosed as a salivary gland choristoma.

Three months later the child was readmitted for further remo-

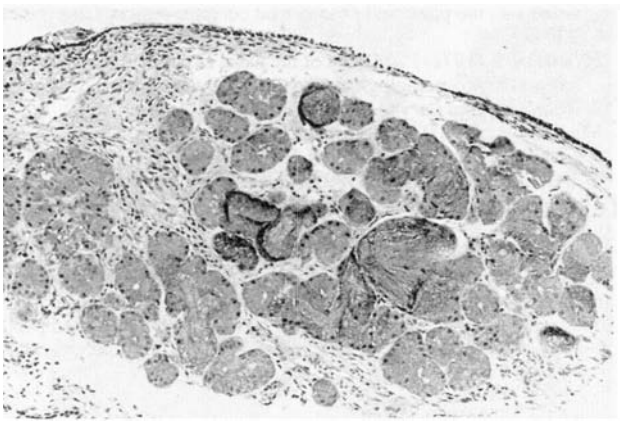

Fig. 1
Low power histology of the initial biopsy specimen showing a small collection of salivary-type acini. (H&E ×100)

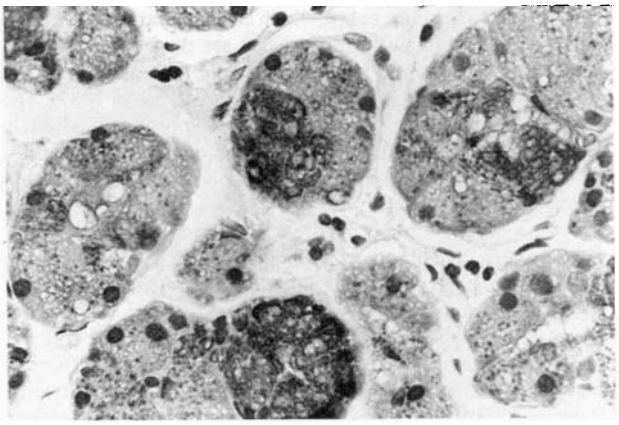

Fig. 2
High power photomicrograph from final resection showing seromucinous acinar structures. (H&E ×250)

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TABLE I
ABNORMALITIES PRESENT AT SURGERY

Malleus	Normal	8	*Incus*	Absent	5	*Stapes*	Not seen	5
	Fused to incus	3		Hypoplastic	1		Normal	1
	Fixed	1		Eroded	6		Abnormal	
	Eroded	1		Inferior site	2		superstructure	8
	Fibrosis around	1		Fixed	1			
	Hypertrophic	1		Normal	2			
	N.C.	8		N.C.	1			
VII Nerve	Abnormal course	5	*Oval W*	Normal	4	*Round W*	Normal	4
	Dehiscent	7		Absent	3		N.C.	9
	Normal	2		Not seen	3		Absent	1
	Post-op palsy	3		Bridged by fibrous tissue	1			
Muscles	No stapedius	4	*Surgery*	Biopsy only	5	*Misc.*	Persistent stapedial art.	1
	No t. tympani	3		Total removal	8		Tumour in mastoid antrum	1
				Tympanoplasty	3		Deformed pinna + unilat facial hypoplasia	2
							Abnormal inner ear	1

N.C.—not commented
W.—window
Misc.—miscellaneous

val of the lesion. This was performed via an endaural approach. The mass peeled away easily to reveal a distorted stapes: the footplate and posterior crus were all that remained. The Fallopian canal was intact and in the normal anatomical position. No attempt at ossiculoplasty was undertaken because of the rare nature of the pathology. We preferred a two stage procedure which would in addition allow assessment of any recurrence at the second operation. At the end of the procedure the facial nerve was intact.

Post-operatively the child made a good recovery, and to date has no sign of recurrence on examination in outpatients. A pure tone audiogram performed two months post-operatively, showed only a 20–30 dB loss despite no attempt at ossiculoplasty.

Discussion

The first reported case of middle ear choristoma was in 1961 by Taylor and Martin. However, Uchytil, in 1956, reported a case of 'cylindroma of the tympanic cavity' which some authors have interpreted as possibly representing a choristoma (Moore *et al.*, 1984).

Of the 18 definite cases reported the ages ranged from 5–52 years with a male to female ratio of 1:1.3. One case is included as a personal communication in the paper by Caplinger and Hora (1967). One case had bilateral choristomas (Peron and Schuknecht, 1975). The most common mode of presentation is with mild hearing loss in patients who are otherwise asymptomatic; the hearing loss varied from 15–70 dB.

The choristoma is often associated with other abnormalities and may constitute a syndrome complex. The most commonly affected structures are the incus and the course of the facial nerve; however as can be seen from Table I, a wide variety of abnormalities of the middle ear and pinna have been recorded. However, Kley (1979) found no abnormality of the middle ear in his case. The nature of these abnormalities has been ascribed to an abnormality of development.

Debate over the embryological origins of the incus and malleus have occurred. The most widely accepted theory currently is that the head of the incus and body of the malleus arise from the first branchial arch mesoderm and that the manubrium of the malleus and the long process of the incus along with the stapes superstructure arise from the second. It is also known that the canal wall of the horizontal portion of the Fallopian canal arises from Reichert's cartilage (Anson, 1960).

It follows that the most commonly seen effects on the long process of the incus, stapes and tympanic portion of the facial nerve (Table I) could all be explained by abnormalities of the second branchial arch development prior to the fourth intrauterine month. In addition, this may account for the rare absence of the tensor tympani (Bruner, 1970) and persistent stapedial artery (Nogura and Haase, 1967).

This theory does not explain some of the other features seen such as a deformed pinna, facial hypoplasia and inner ear abnormalities as reported by Peron and Schuknecht (1975). Therefore in its greatest extent, one must theorize abnormal development of the first and second arch, auricular tubercles and the primitive otocyst.

The origin of the middle ear choristoma remains obscure. Peron and Schuknecht propose that the salivary tissue develops from trapped ectodermal rests as in congenital cholesteatoma. Hociota and Ataman (1975) feel that these anomalies may arise from aberrant parotid budding the presence of which subsequently stimulates morphological and functional differentiation of the surface epithelium.

Batsakis (1989) reviewed tumours of the middle ear and divided them into three groups:

1. Secondaries from adjacent tissue.
2. Tumours or choristomas of salivary origin.
3. Primary adenomatous tissue *e.g.* adenocarcinoma.

In addition, he reports heterotopic salivary tissue to be found in pituitary, external ear, thyroglossal duct, mandible, and neck (Batsakis, 1986).

By virtue of the rarity of choristomas of the middle ear, there is no established management plan. In the cases reported so far, treatment was dictated by the extent of the lesion and the confidence of the authors in identifying the anatomy of the area as this can often be abnormal. In some cases the tumour is attached via a fine stalk and hence facilitates easy removal (Caplinger and Hora, 1967; Cannon, 1980; Quaranta, 1981).

Biopsy only was performed in five patients (Taylor and Martin, 1961; Steffen and House, 1962; Mischke *et al.*, 1977; Wine and Metcalf, 1977; Kartush and Graham, 1984) because the tumour was attached to the facial nerve and it was felt removal may lead to facial nerve damage. In one case removal was aborted due to the onset of vertigo during a local anaesthetic procedure (Kartush and Graham, 1984).

In all other cases complete removal was possible. Tympanoplasty was attempted in four patients (Nogura and Haase, 1964; Bruner, 1970; Saeed and Bassis, 1971; Abadir and Pease, 1978) with good results in three.

In four cases post-operative weakness of the facial nerve was recorded (Taylor and Martin, 1961; Bruner, 1970; Wine and Metcalf, 1977; Hociota and Ataman, 1975) with full recovery occurring in three cases and a partial recovery in one (Hociota and Ataman, 1975).

Follow-up has shown slight or no growth in periods up to 17 years.

Conclusion

Evidently this is an extremely rare tumour and unlikely to be considered in the pre-operative differential diagnosis. Its diagnosis will usually be made at operation and raises the question of what should be done. From previous cases it appears to be extremely slow growing as indicated by the case of Hociota and Ataman (1975). The proximity and abnormality of the facial nerve would support a policy of biopsy only and observation. If however the lesion is obviously polypoid it may be easy to excise it at its stalk.

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