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Aggressive papillary tumour of the nasopharynx followed by an aggressive papillary tumour of the middle ear. A multiple site tumour?

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Abstract
We report the case of a 72-year-old male presenting with a papillary adenocarcinoma of the middle ear. He had had a similar tumour excised from the ipsilateral nasopharynx seven years previously with no evidence of local recurrence. We conclude that this middle ear lesion possibly represents a second tumour. No record of such a case has been reported previously in the literature. We discuss presentation and management and highlight the need for close follow-up of these patients.

Key words: Adenocarcinoma, Papillary; Ear, Middle; Nasopharynx

Introduction

Middle-ear papillary adenocarcinoma is a rare malignant, locally invasive neoplasm arising from the middle-ear mucosa, associated with destruction of the temporal bone.

Case report

A 72-year-old man was referred from his general practitioner to the ear nose and throat out-patient clinic with a six months' history of constant left serosanguinous otorrhoea. There was no tinnitus or vertigo. A hearing aid user, he had noticed a recent deterioration in his hearing. Seven years previously he had had an ipsilateral nasopharyngeal papillary adenocarcinoma (Figure 1) removed via a Le Fort I down fracture of maxilla and had remained well with no evidence of recurrence. He had not received radiotherapy at that time.

Examination showed the right ear to be normal. On the left a considerable amount of blood and mucus, which was obscuring the tympanic membrane, was micro-suctioned revealing a large polypoidal mass filling the medial aspect of the left external acoustic meatus. Examination of the cranial nerves, nasopharynx, oral cavity, larynx and neck was normal.

A computed tomography (CT) scan of the skull base showed material of soft tissue density throughout the left middle ear and into the mastoid air cells. Magnetic resonance imaging (MRI) of the neck showed no evidence of tumour within the nasopharynx or extension to adjacent soft tissues or into the left middle ear. The patient underwent an urgent examination under general anaesthesia and biopsies were taken of his left middle ear and nasopharynx. The histology of the middle-ear lesion showed a neoplasm with a papillary-glandular appearance suggesting a low-grade papillary adenocarcinoma (Figures 2 and 3) very similar to the lesion that had been removed from the nasopharynx seven years previously. The sections

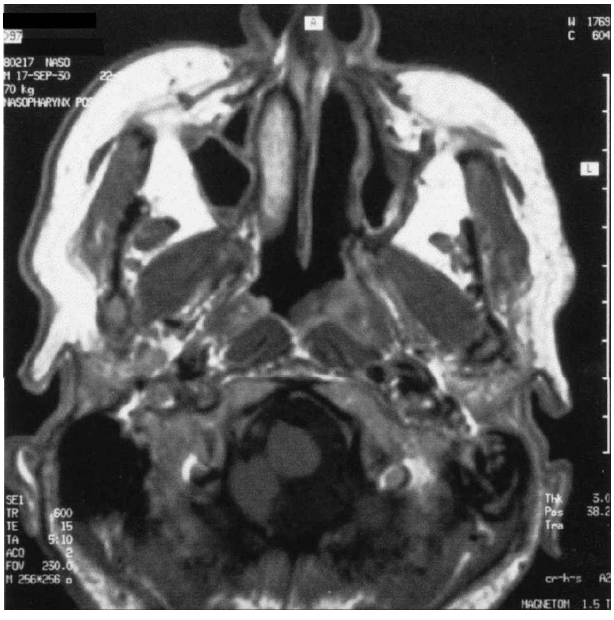

Fig. 1
MRI of nasopharynx showing a lesion of the left nasopharynx with normal tissue between the mass and the lateral pterygoid muscle.

from the recent nasopharyngeal biopsy did not show any evidence of malignancy. Based on the patient's age, pathology report, previous history and the obvious extensive involvement of left temporal bone, we proceeded to a debulking of the tumour in the form of a radical mastoidectomy followed by radical radiotherapy. The patient made an uneventful recovery and remains under regular review.

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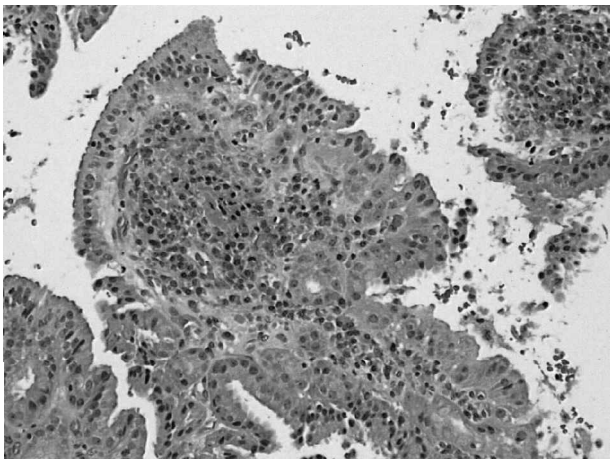

FIG. 2

The tumour has a complex papillary architecture with some entrapped glands in fibrous stroma (H & E; ×40)

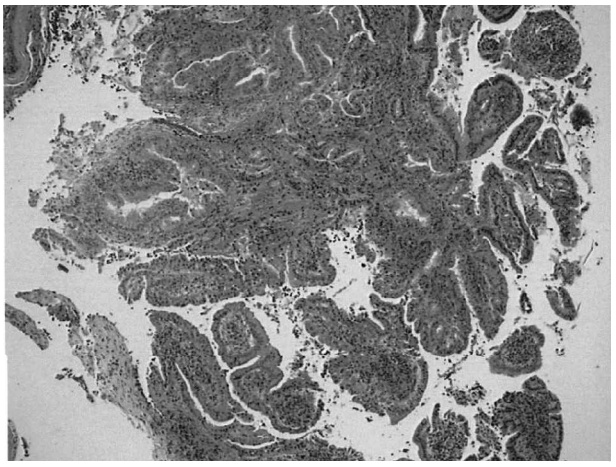

FIG. 3

On higher magnification the papillae are lined by a single layer of eosinophilic columnar epithelial cells with minimal nuclear pleomorphism lining chronically inflamed fibrovascular stroma (H & E; ×160).

Discussion

In this case report there are two important focusing points: firstly, the possible oncogenic origin of this tumour at multiple sites and secondly, the need for close follow-up of patients with this type of tumour.

- **Case of a 72-year-old male presenting with a papillary adenocarcinoma of the middle ear**
- **A similar tumour was excised seven years previously from his ipsilateral nasopharynx with no evidence of local recurrence**
- **The presentation, management and need for close follow-up are discussed**

'Aggressive papillary tumours' of the middle ear are rare locally invasive and destructive malignant neoplasms. Previously known as papillary adenocarcinomas, newer evidence suggests they arise from the endolymphatic sac.[1] The commonest symptoms are purulent or serosanguinous otorrhoea, otalgia and hearing loss.[2] Other symptoms include vertigo, tinnitus (often pulsatile) and aural fullness. Tumour progression can lead to facial nerve weakness and other cranial nerve involvement.[3] Some patients will give a long history of otological symptoms, including ear operations and occasionally a history of recurrent childhood ear infections.[2] Despite their locally invasive character they do not appear to metastasize.[3] Radiological studies of these tumours usually show soft tissue densities with, or without, temporal bone destruction depending on the aggressive nature of the tumour.[3,4] These lesions may lie 'between the sigmoid sinus and the internal auditory canal' or they can invade the posterior fossa.[5] Heffner was the first to add the endolymphatic sac's epithelium as a possible origin for the papillary neoplasms of the ear.[6] There are two histological groups of adenomatous tumours involving the middle ear: mixed and papillary.[7] The mixed type has an acinar pattern of glandular organization, is usually confined to the middle ear and mastoid and often presents as chronic suppurative otitis media.[1] The papillary type demonstrates villous formation and usually presents late after local tissue destruction.[1] Facial nerve involvement is uncommon in mixed patterns, whereas papillary tumours involve the facial nerve and may extend to the petrous area and the middle or posterior fossa.[1] With respect to the differential diagnosis, other ear tumours, apart from the aggressive papillary middle-ear tumours, need to be considered. Jugulotympanic paragangliomata (chemodectoma, glomus jugulare tumour, glomus tumour) arise in the jugular bulb, are locally invasive and have a specific histological pattern including neuronal differentiation without glandular structures.[8] Other tumours such as basal cell or squamous cell carcinomas and malignant ceruminomas may extend locally and intracranially.[8]

The terminology applied to adenomatous tumours of the temporal bone is confusing and reflects the difficulty in diagnosis of these lesions due to overlapping pathological features and the necessity for close collaboration between surgeons, pathologists and radiologists.[9] It seems that the more modern term 'endolymphatic sac tumour' is here to stay.

The origin of these destructive tumours remains undefined. Hyams and Michaels re-examined some of their own cases originally thought to be primary adenocarcinomas and revised their diagnosis with the tumours being designated as metastatic from sites such as postnasal space, breast and parotid gland.[10] The question of whether such tumours arise from neural crest cells in the middle ear is as yet unanswered.

Treatment of a middle-ear papillary adenocarcinoma involves local resection of the tumour or radical mastoidectomy alone, or in combination with, radiotherapy. In advanced cases the latter alone may be the sole option,[2] although there are no studies focusing on the role of adjuvant radiotherapy.

Despite the fact that distant metastases have not been reported in the literature, the rarity of papillary adenocarcinomas may justify a routine metastatic workup.[11] Our patient presented with two histologically identical tumours; a papillary adenocarcinoma of the nasopharynx followed – seven years later – by a middle-ear papillary adenocarcinoma. Our first thought was that the eustachian tube was the route of tumour spread but biopsies of the area were negative. There is no history of radiotherapy in the first tumour, therefore it could be argued that this represents a second tumour. We cannot decide whether the primary originated from the nasopharynx or the ear or both. A close follow-up review of patients with this type of tumours is necessary due to the possible oncogenic origin of this tumour at multiple sites.

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