Challenges in diagnosing ceruminous adenocarcinoma

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DESCRIPTION

A 65-year-old man presented to the Ear, Nose and Throat (ENT) clinic with right-sided otalgia, discharge and non-healing ulcer on the outer ear canal for a couple of months. His medical history includes diabetes and chronic obstructive pulmonary disease. On examination, he had a subcentimetre ulcer in the right mid-ear canal. The rest of the ENT examination was normal.

Initial biopsy of the ear canal ulcer was non-diagnostic although atypical cells were noted. A repeat biopsy showed features compatible with basal cell carcinoma (BCC). CT and MRI scans of the neck showed 2 cm soft tissue mass in the right ear canal extending to the adjacent parotid gland (figure 1). The patient had limited lateral petrosectomy and superficial parotidectomy to achieve primary clearance. Postoperative histology showed ceruminous adenocarcinoma (CA) with positive margins. A further resection was planned, but unfortunately, the patient died from causes unrelated to the primary ear lesion.

Ceruminous glands are modified apocrine adnexal glands of the external auditory canal (EAC) and rarely gives rise to malignant changes. There are few natural barriers in the EAC; malignant tumours can spread into the parotid gland and surrounding lymphatics or to the temporal bone. CA poses a challenge to pathologists as this is a rare tumour, with one large centre reporting only 0.00025% of all the surgical specimens, and 2.4%–5% of all ear canal malignancies.^{1,2} Histologically, the lesion was a highly infiltrative basaloid tumour with cribriform, tubular and cord-like infiltrative patterns with perineural invasion (figure 2A–C). It was neither originating from the epidermis of the ear canal nor from the parotid. The morphology was that of an adenoid cystic carcinoma, and with origin from the ear canal, it was compatible with a CA, adenoid cystic carcinoma subtype.

Figure 2 Histology of specimen showing highly infiltrative basaloid tumour with (A) cribriform, (B) tubular and (C) cord-like infiltrative pattern with perineural invasion (red arrow) compatible with ceruminous adenocarcinoma (H&E, original magnification $\times 200$).

The initial punch biopsy was reported as a BCC; the diagnosis was revised following examination of the larger excision specimen. This highlights the potential pitfall of a small biopsy of a rare tumour, CA, which partly resembles a much more common tumour BCC. However, histopathological analysis includes both incisional and excisional specimens and a diagnosis can occasionally change with assessment of more tumour material.

CA has non-specific symptoms including otalgia, discharge, ear fullness, vertigo, tinnitus, hearing loss, ear bleeding, ear canal polyp and facial palsy, which could initially suggest a simple benign inflammatory condition.^{3,4} Considering this patient is diabetic with disproportionate otalgia, necrotising otitis externa was initially entertained. CA can present as prolonged subclinical phase lasting for years, with mean duration of symptoms in literature at 51.6 months.^{1,5} It has been reported to spread to lungs, bone, liver and kidneys, but less on neck nodes.

The primary treatment of CA is combined surgery and postoperative radiation.⁴ Primary surgical clearance poses technical difficulties and could include wide en bloc excision, parotidectomy and neck dissection. Perineural invasion is common and recurrence is as high as 90% if parotid gland was initially involved.¹ Even with negative margins following resection, recurrence and distance

Figure 1 Axial and coronal views of MRI neck showing 2.0xcm1.6 cm soft tissue mass (red arrow) beneath the right ear canal extending to the adjacent parotid gland.

Learning points

- ▶ A wax-producing gland in the ear canal can give rise to a neoplastic process, which can be benign or malignant.
- ▶ Ceruminous adenocarcinoma (CA) is rare and can pose a diagnostic challenge to pathologists in small biopsies.
- ▶ The symptoms of CA are very non-specific and are similar to benign inflammatory lesions; it has a prolonged subclinical phase for years.

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metastasis occur.⁶ On average, patients with CA had a mean time from initial presentation to death of 4.7 years.¹

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