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Clinical Records

Pilomatrixoma of the external auditory meatus

Bippon C. Vinayak, F.R.C.S., Graham J. Cox, F.R.C.S., Margaret Ashton-Key, M.B.*

Abstract
Pilomatrixoma is a benign skin tumour arising from the outer hair root matrix. It is an uncommon tumour which favours the hair bearing area of the head and neck. The preauricular area is a well documented site, but to our knowledge, there are no reports of a pilomatrixoma arising in the external auditory meatus. We present such a case and discuss the historical background along with its pathological and clinical features.

Key words: Ear, external; Ear neoplasms; pilomatrixoma

Case report

A nine-year-old Caucasian boy first presented to his General Practitioner with a three-week history of a painless lesion in the left ear canal. It was treated initially with antibiotics and then with silver nitrate cautery. However, it enlarged and became painful. He was, therefore, referred to the ENT out-patient department where he was found to have a 5 mm firm, polypoidal lesion arising from the anterior wall of the outer ear canal (Fig. 1). The overlying skin was intact and had a dusky red colour. It was tender but did not bleed on contact. The remainder of the otological examination was normal. Clinically, this lesion was thought to be a pyogenic granuloma. Excision biopsy was performed. Histology showed a normal epidermis, but within the dermis there was a well circumscribed tumour composed of islands of 'basophils' and of 'shadow' cells. The surrounding stroma contained multinucleated giant cells, areas of keratanization and calcium deposits (Fig. 2). the features were considered classical of a pilomatrixoma.

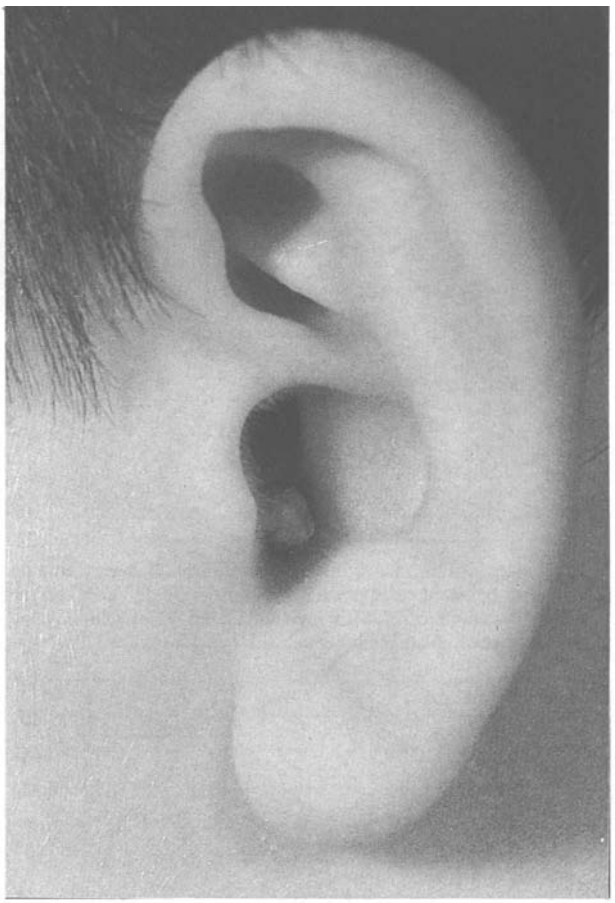

Fig. 1
Pilomatrixoma of the left external auditory meatus.

Discussion

In 1880, Malherbe described a calcified tumour originating from the sebaceous gland and it subsequently acquired the eponym, 'calcifying epithelioma of Malherbe' (Malherbe and Chemantais, 1880). Forbes and Helwig studied 228 such tumours in 1961 and renamed it 'pilomatrixoma'. This was based on histochemical and electron microscopic evidence, which established the outer root sheath cell of the hair follicle as the cell of origin. The estimated incidence is approximately 1 in 1000 skin biopsies. At presentation, over 60 per cent of patients are under 20 years old, with the majority (two-thirds) occurring in children under 10 years of age (Moehlenbeck, 1973). In the literature a 3:2 female:male predominance is often quoted, although several authors report an essentially equal sex distribution (Moehlenbeck, 1973; Makek *et al.*, 1989). Pilomatrixoma often occurs in sites where a dermoid or inclusion cyst is found, such as the preauricular region, eyebrow and external angle of the eye. These other lesions may form the differential diagnosis and has led to the suggestion that a pilomatrixoma has an

From Department of Otolaryngology, Radcliffe Infirmary, Woodstock Road, Oxford OX2 6HE, and Department of Histopathology*, John Radcliffe Hospital, Oxford.
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embryological origin arising from degeneration of an embryological hair follicle cyst (MacLeod and Scobie, 1991). The head is involved in approximately 57 per cent of cases (eyebrow 14 per cent, cheek 16 per cent, preauricular 16 per cent and forehead 11 per cent), the upper limb is affected in 20 per cent of cases whilst the neck is involved in 11 per cent of cases. The remaining cases generally occur on the upper trunk, other sites being rarely affected. Histologically, there is differentiation towards hair cortex cells. Generally, it is a well demarcated lesion which lies in the dermis with extension into subcutaneous fat. It is composed of two main types of epithelial cells: 'Basophilic cells' having deeply basophilic nuclei, lying close together with scant cytoplasm, and 'shadow' or 'ghost' cells with a distinct border and a central unstained area which is the 'shadow' of the lost nucleus. This cytological appearance is unique to a pilomatrixoma. As the lesion ages, there is a transition from the 'basophilic' cells to the 'shadow' cells. The lesion may be surrounded by fibrosis due to a foreign body giant cell reaction to keratin. Calcium deposits devoid of lamellations may also be present.

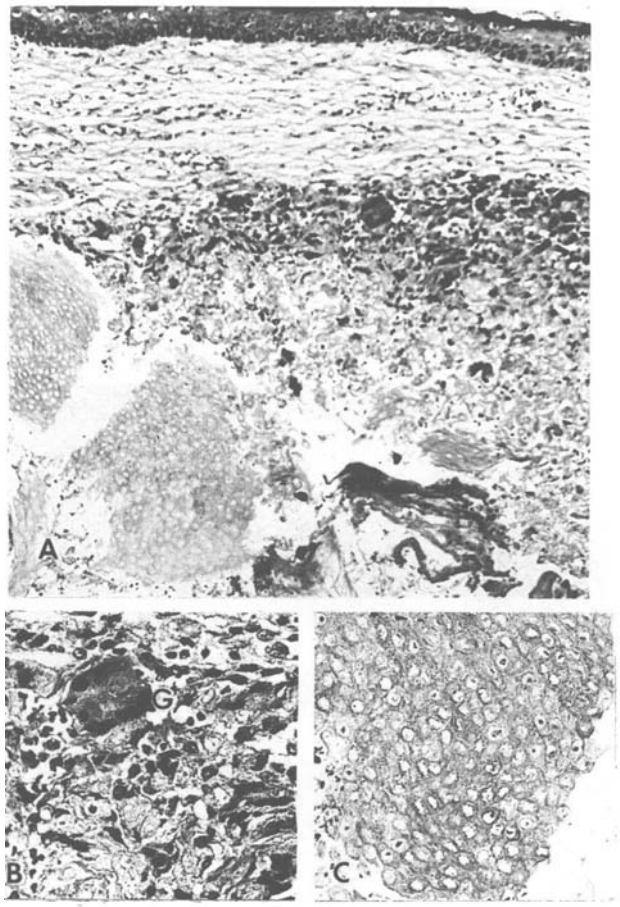

FIG. 2

(A) Low power of pilomatrixoma with epidermis above and tumour below, with basaloid cells (right) and ghost cells (left). (B) Higher power of basaloid cells with multinucleated giant cell (G). (C) Higher power of ghost cells.

Clinically, a pilomatrixoma presents as a solitary, firm and well circumscribed lump in the dermis or subcutaneous tissue. Multiple tumours occur in 2 per cent to 3 per cent of cases. The overlying skin may show a bluish discolouration but there are no distinctive clinical features. The differential diagnosis includes sebaceous cysts, dermoid or inclusion cysts, calcified haematoma, haemangioma and lymph node (Hawkins and Chen, 1985). The diagnosis is usually made histologically having been overlooked clinically.

Treatment of this benign tumour is local excision. However, in 1980 Lopansri and Mihm described a malignant variant and some of these tumours are believed to have arisen in pilomatrixoma. So far, no malignant cases have been reported in children and distant metastases have not been seen.

A pilomatrixoma needs to be borne in mind when considering the differential diagnosis of a lesion in the outer ear canal. Local excision is recommended as the treatment of choice.

Acknowledgement

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Address for correspondence:
Mr Bippon C. Vinayak, F. R.C.S.,
Department of Otolaryngology,
Radcliffe Infirmary,
Woodstock Road,
Oxford OX2 6HE.