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Clinical Records

Primary amyloidosis of the external auditory canal: case report

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Abstract
A case of primary amyloidosis of the external auditory canal is described. To the best of our knowledge this is the first case described in the literature.

Key words: Ear canal; Amyloidosis

Introduction

Amyloidosis refers to the accumulation in the tissues of various insoluble fibrillar proteins (amyloid substance). There are several classifications but in general four clinical forms are recognized (Kisilevsky, 1988): (1) primary or idiopathic; (2) secondary or reactive; (3) hereditary; and (4) localized.

Primary localized cutaneous amyloidosis designates amyloidosis of the skin in the absence of systemic involvement and has been classified into five types: nodular, macular, lichen and poikiloderma-like amyloidosis, familial primary cutaneous amyloidosis (Hicks *et al.*, 1988).

There have been various reports of primary localized amyloidosis of the auricle but to the best of our knowledge no reports of a case affecting the external auditory meatus.

Case report

A 63-year-old man was referred to the ENT outpatient clinic at the Royal Liverpool University Hospital in April 1991 with a three-year history of intermittent right otalgia and blood-stained otorrhoea. The patient had no previous history of ear disease and physical examination showed a granular lesion of the postero-superior wall of the right external auditory canal, 2 mm lateral to the tympanic membrane. It was felt that the lesion could be a neoplasm and a biopsy was taken. The lesion was later excised through a post-auricular approach under general anaesthetic.

On haematoxylin and eosin staining the specimen showed stratified squamous epithelium with nodules of eosinophilic material in the dermis, there was no evidence of a neoplasm. The eosinophilic material showed the characteristic birefringence of amyloid when stained by Congo Red and viewed through crossed polarizing filters (Figure 1).

CT scan of the external auditory canal and mastoid did not show any evidence of bony destruction or invasion. There was no evidence of systemic amyloid disease. Full blood count, erythrocyte sedimentation rate, protein electrophoresis, plasma chemistry, urine proteins, chest X-ray, electrocardiogram, ultrasound scan of liver and spleen were all normal.

At follow-up a year later, the patient was well and asymptomatic.

Discussion

We have described a case of primary localized cutaneous amyloidosis of the external auditory canal. In the literature 12 cases of primary, localized amyloidosis of the auricle have been described (Sanchez, 1983; Dupre *et al.*, 1984; Weissbluth *et al.*,

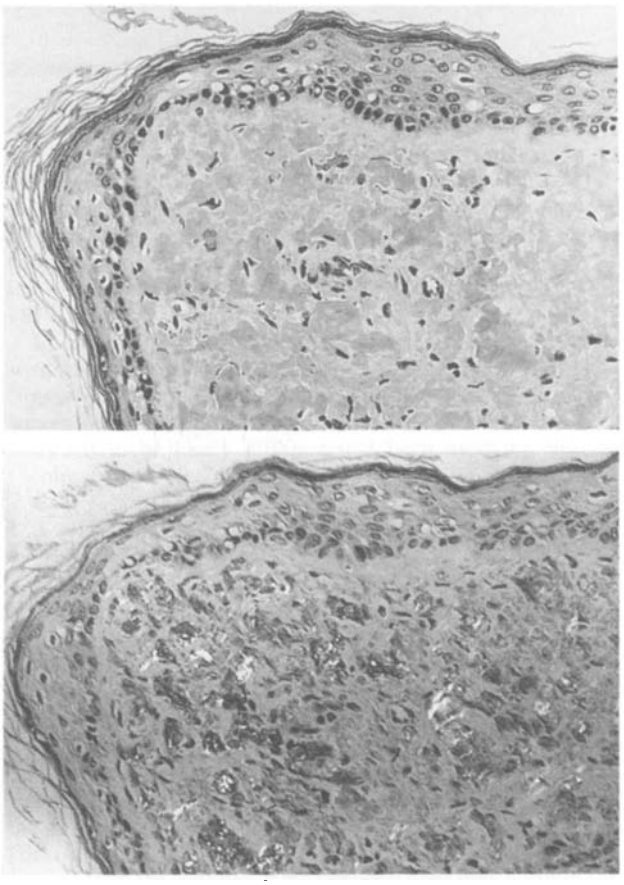

FIG. 1

Skin from the external auditory meatus showing amyloid deposits in the dermis. (a) Stained with H & E; (b) Stained with Congo Red (viewed through crossed polarizing filters). (×280)

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1987; Hicks *et al.*, 1988; Barnardas *et al.*, 1990), but to the best of our knowledge this is the first report of a lesion involving the external auditory canal.

Giordano *et al.* (1981) describe a case of amyloidoma of the temporal bone with invasion of the external auditory canal, but this distinctive entity is a benign tumour producing amyloid for which local excision and radiotherapy have been advocated (Lipper and Khan, 1978).

The importance of our observation is that the symptoms and appearance of the lesion raised suspicion of a neoplasm. Localized amyloidosis of the external auditory canal should be considered in the differential diagnosis of granular lesions associated with blood-stained otorrhoea and otalgia. Local excision of such a lesion appears to be curative.

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