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Clinical Records

Juvenile xanthogranuloma auris: an atypical presentation

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Abstract
We present a case of juvenile xanthogranuloma of the external ear canal in a 51-year-old woman and a review of the literature.

Key words: Ear neoplasms; Ear canal; Xanthogranuloma, juvenile

Introduction

In 1905 Adamson was the first to describe juvenile xanthogranuloma in the English literature with the name of xanthoma multiplex (Adamson, 1905). The alternative name juvenile xanthogranuloma (JXG), proposed by Helwig and Hackney (1954) is now in use. Two-thirds of cases present within the first nine months of life, but late onset in adolescence or adult life occurs in a small percentage of cases. There does not appear to be any familial or racial predisposition, and no concomitant abnormalities of lipid metabolism.

The lesions usually present as solitary or multiple papulo-nodules (1–10 mm in diameter) and are most frequently found on the head and neck, upper part of the trunk and proximal parts of the limbs (Weedon, 1992). In occasional cases there may be ocular or visceral involvement. Atypical forms with extensive facial or generalized eruptions have also been reported.

Case report

A 51-year-old Asian woman presented with mild conductive deafness and a small nodule in the left ear canal. She was not taking any medication and had no known allergies. There was no significant past medical history other than vitiligo. Examination of the left ear revealed a 1.1 × 0.2 × 0.1 cm polypoid lesion arising from the postero–superior aspect of the meatus. The lesion was removed under general anaesthesia. Post-operatively she was well and is currently asymptomatic.

Pathological findings

The morphology of the lesion, as seen with the light microscope, conformed to the well-described pattern of juvenile xanthogranuloma (Figure 1).

The polypoid growth showed richly cellular stroma covered by stratified squamous epithelium. The proliferating cells were histiocytes with foamy cytoplasm, involving the dermis and extending up to, but not invading the epidermis. The cells were polygonal or spindle-shaped with indistinct cytoplasmic borders, and showed no atypia. Scattered among the histiocytes were Touton-type ('wreath-like') and foreign body-type giant cells. A moderate degree of interstitial fibrosis was seen throughout the entire lesion. Blood vessels showed some endothelial swelling. A scattering of inflammatory cells was present, including occasional eosinophils.

Discussion

The clinical picture of this case is unusual as this lesion has not been described in the ear canal before and it most commonly occurs at an earlier age. The histopathology was however typical of juvenile xanthogranuloma (JXG).

Juvenile xanthogranuloma must be differentiated histologically from many other conditions such as cutaneous histiocytosis X. There are fewer eosinophils in JXG; it does not generally invade the epidermis; and there is greater cellular cohesion than in histiocytosis X (Enzinger and Weiss, 1992). Xanthomas consist of a more uniform population of foam cells and contain fewer Touton-type giant cells. Furthermore, large extracellular cholesterol deposits can be seen in xanthomas associated with hypercholesterolaemia.

Reticulohistiocytoma is characteristically composed of a collection of large multinucleate cells with randomly arranged nuclei and glassy cytoplasm. Benign fibrous histiocytoma is distinguished from JXG by the predominant storiform appearance, thick collagen fibres and hyperplastic epidermis. In JXG a storiform pattern, if present, is indistinct and the covering epidermis is often thin. JXG, especially those located in deep tissues, may be mistaken for a malignant tumour such as malignant fibrous histiocytoma or embryonal rhabdomyosarcoma. JXG shows little nuclear pleomorphism and lacks rhabdomyoblasts (Sonoda *et al.*, 1985).

A report by Tahan *et al.* (1989) states that JXG shows several recognizable histological patterns; xanthogranulomatous, xanthomatous, fibrohistiocytic, or combined. In view of the known natural history of spontaneous involution, it is possible that these variants are attributable to evolutionary stages of a lesion.

The question remains as to whether these lesions

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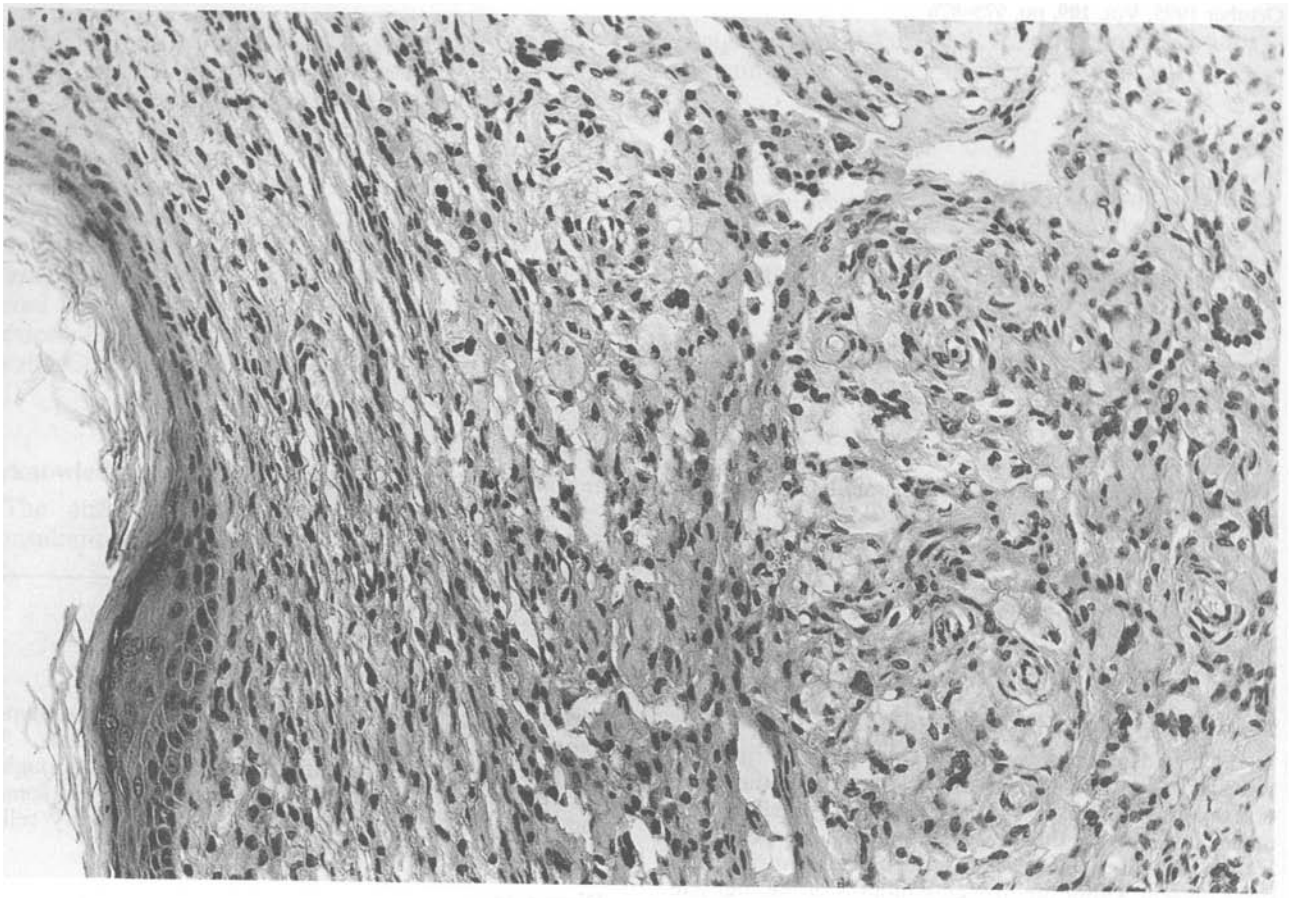

FIG. 1
Microscopy of juvenile xanthogranuloma showing Touton-type giant cells amongst foamy histiocytes. (H&E; ×200).

represent a true neoplasm or an unusual reactive process (Enzinger and Weiss, 1992). The involutionary nature of the disease, especially in children argues for a reactive process. A case of a JXG occurring in a parotid gland with associated cytomegalovirus suggests that the disease may represent a response to viral infection. Indeed localized histiocytic 'tumours' can be induced experimentally, or occur spontaneously in monkeys following viral infection. They are composed of a proliferation of histiocytes and eventually regress (Enzinger and Weiss, 1992).

The prognosis of the lesions is very good. Most adult patients in documented studies had a solitary lesion that was removed and did not recur. In children the skin lesions ultimately regressed and even deeply located tumours pursued a favourable course (Enzinger and Weiss, 1992). Lesions in the eye have an associated danger of haemorrhage into the anterior chamber and consequent glaucoma. In such cases radiotherapy has been employed successfully (Webster *et al.*, 1966). Radiotherapy has also been used for lesions in less accessible locations such as the pericardium.

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