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Angiolymphoid hyperplasia with eosinophilia

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Abstract
Angiolymphoid hyperplasia with eosinophilia is a rare benign condition that causes swellings in the head and neck. It is difficult to diagnose prior to biopsy and is frequently mistaken for a malignant tumour. A case involving a 21-year-old man who presented with a 2 cm diameter fibrous lesion in the subcutaneous tissue of the cheek is reported. The clinical and histological features are reviewed and the differences between this condition and the similar condition of Kimura's disease are discussed. Initial treatment with intralesional or systemic steroids is suggested as this may avoid the need for excision.

Key words: Angiolymphoid hyperplasia with eosinophilia; Cheek

Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare cause of swellings in the head and neck. It was first described in 1969 in a report of nine patients with persistent subcutaneous nodules who presented to a dermatology department (Wells and Whimster, 1969). Subsequent reports have shown the predilection of the disease for the head and neck (Olsen and Helwig, 1985) and thus all surgeons involved in treating conditions in this area should be aware of the clinical features and management options available.

Case report

A 21-year-old Malaysian man presented with a 2 cm diameter mass in his right cheek, lying deep to the skin and anterior to the parotid region. It had slowly increased in size over two years. There was no cutaneous or mucosal ulceration. The mass was firm, had a smooth surface and was not tender. There were no palpable lymph nodes. A full blood count showed a white cell count of 8.1×10^9/l with an eosinophilia of 1.76×10^9/l (0.04–0.44×10^9/l normal range). Fine needle aspiration cytology was obtained, and showed no malignant cells, but demonstrated loose connective tissue, lymphocytes, macrophages and eosinophils. A CT sialogram showed the mass to be separate from the parotid gland, lying in the subcutaneous tissues.

The lump expanded rapidly to 6 cm diameter over the subsequent three weeks. In order to exclude the possibility of malignancy, the mass was excised via a skin crease incision. A well vascularized fibrotic mass was found to extend to within 5 mm of the oral mucosa. It involved small branches of the facial nerve, which could not be dissected free and had to be sacrificed. Postoperative recovery was good, and there was no clinical impairment of facial nerve function.

Histological examination showed a proliferation of small blood vessels lined by plump endothelial cells and surrounded by a lymphoid infiltrate with formation of germinal centres. Large numbers of eosinophils were seen (Figure 1). A diagnosis of ALHE was made.

Discussion

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare condition of uncertain aetiology. There are wide racial differences in presentation, with oriental patients being predominantly male (85 per cent) and young (second decade) in contrast to western cases which tend to occur in females (70 per cent) and older patients (third or fourth decades) (Henry and Burnett, 1978). It causes papular or nodular angiomatous lesions in the dermis, subcutaneous tissues and adjacent lymph nodes, which average 1 cm in diameter (Kung *et al.*, 1984). Distribution is almost entirely restricted to the head and neck (Olsen and Helwig, 1985) and there seems to be a predilection for the area around the external ear and external auditory canal (Vallis and Davies, 1988; Murty and Cox, 1990; Sharp *et al.*, 1990) although there are reports involving the orbit (Smith *et al.*, 1988), lacrimal gland (Cook and Stafford, 1988), oral mucosa (Buckerfield and Edwards, 1979) and the arm (Akosa *et al.*, 1990).

Originally ALHE was thought to be equivalent to Kimura's disease, a condition prevalent in Japan, China and South-east Asia which also causes angiomatous skin lesions, together with lymphadenopathy and marked eosinophilia (Kimura *et al.*, 1948), and the terms have often been used synonymously (Smith *et al.*, 1988). However recent clinicopathological studies have suggested that these are two separate conditions which have different clinical and histological features (Rosai *et al.*, 1979; Googe *et al.*, 1987; Urabe *et al.*, 1987). These authors suggest that ALHE represents a stage of histiocytoid or epithelioid haemangioma, which is a true vascular neoplasm, whereas they view Kimura's disease as a localized manifestation of a systemic immunological reaction. Other authors disagree that ALHE is a vascular neoplasm; they believe that the entity represents a localized atopic reaction to a variety of agents (Hallam *et al.*, 1989; Akosa *et al.*, 1990). It therefore needs to be differentiated from epithelioid haemangioma, which has similar histological features to ALHE regarding endothelial cell morphology, but is without the eosinophilic infiltrate or formation of germinal centres which suggest an immunological reaction. Our case had a marked eosinophilic infiltrate and is therefore unlikely to represent an epithelioid haemangioma.

The appearances of the lesions in ALHE and Kimura's disease are similar, with a raised erythematous skin lesion or a fibrous subcutaneous nodule being common. Histological examination of ALHE shows exuberant proliferation of blood vessels lined by plump endothelial cells as seen in this case. Such vessels may

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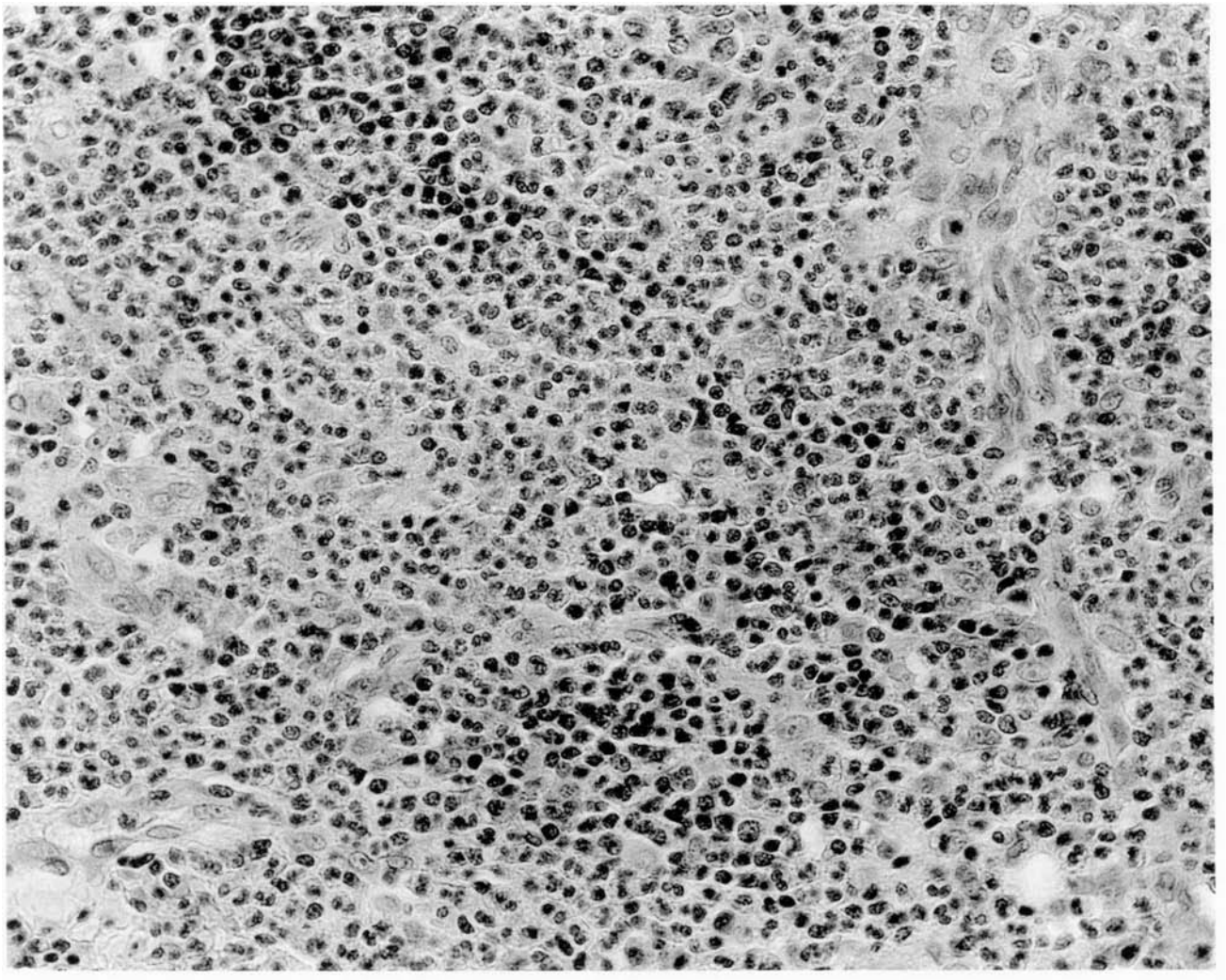

FIG. 1

ALHE: prominent endothelial cells with surrounding lymphoid aggregates and eosinophils (H&E; ×200).

be uncanalized. There is an accompanying perivascular lymphocyte and plasma cell infiltrate, and there may be germinal centre formation. Kimura's disease is more likely to present with a solitary larger mass, and to be accompanied by lymphadenopathy and peripheral eosinophilia. The newly formed blood vessels are canalized, and lined by flat endothelial cells. (Rosai *et al.*, 1979; Kung *et al.*, 1984) (See Table I).

It is difficult to be certain whether our case is an example of ALHE or Kimura's disease; the age, sex and race of the patient support Kimura's disease, but the histological picture is more in favour of ALHE. The exact distinction is unlikely to be of great importance, as these diseases probably represent a similar reactive process in the tissues, with the minor histological differences depending on whether the insult is localized or systemic.

ALHE is benign, and may regress spontaneously, but the majority of masses persist as slow growing tumours (Wells and Whimster, 1969). There are no reports of malignant change. The appearances of the lesion have led it to be mistaken for Kaposi's sarcoma, malignant lymphoma and angiosarcoma, as well as pyogenic granuloma, haemangioma and dermatofibroma (Buckerfield and Edwards, 1979; Barnes *et al.*, 1980) and in one series of 116 cases only one was correctly diagnosed clinically prior to excision (Olsen and Helwig, 1985). This difficulty in diagnosis was also our experience, even though fine needle aspiration

TABLE I

IMPORTANT CLINICAL AND HISTOLOGICAL DIFFERENCES BETWEEN ALHE AND KIMURA'S DISEASE

Clinical	ALHE	Kimura's disease
Anatomical distribution	Predominates in head and neck area. May be multiple. Usually 1–2 cm diameter	Generalized Often solitary May be larger than 2 cm
Lymphadenopathy	Local	Systemic
Peripheral eosinophilia	Less common	Marked
Histological appearances		
New vessels	Exuberant proliferation of new blood vessels, which may be uncanalized, with plump endothelial cells	New vessels canalized, and lined by flat endothelial cells

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cytology showed intralesional eosinophilia. A raised awareness of this disease is important, as accurate clinical and cytological diagnosis may allow for initial treatment with intralesional or systemic steroids instead of excision (Linwong *et al.*, 1990). Surgical excision, perhaps using laser to reduce haemorrhage (Vallis and Davies, 1988) could then be reserved for residual or recurrent masses.

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