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Angiosarcoma of the head and neck

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Abstract
Angiosarcoma of the head and neck is a rare tumour of vascular origin that affects the elderly. A 74-year-old man who presented with bruise-like macules of the scalp and face is reported. He was treated for a few months with different antibiotics and anti-allergic medication by his own family doctor, and referred for specialist opinion when he failed to derive any benefits from the medications. A biopsy was obtained from the lesion and proved to be an angiosarcoma. A review of the literature indicates that the most important prognostic factor in this particular disease is the size of the lesion on presentation, hence the importance of early diagnosis. This case is reported, and the literature for similar cases is reviewed, to highlight the diagnostic and therapeutic aspects of this uncommon aggressive tumour in an attempt to help in the process of early diagnosis.

Key words: Haemangiosarcoma; Chemotherapy; Radiotherapy

Case report

A 74-year-old male presented with pain and bruise-like macules on the scalp and face of a few months' duration, unresponsive to antibiotics and antihistamines.

On clinical examination, he was generally fit and apyrexial. There were extensive bruise-like macules of the scalp and face with induration over the cheek and temple, and chemosis of eyelids (Figure 1). The remainder of the examination was entirely normal.

A full blood count, differential and ESR were within normal ranges. Chest X-ray showed no abnormalities, and a computed tomography (CT) scan to the head and neck showed soft tissue swelling affecting the scalp and extending down to the upper face. Several biopsies obtained from different areas of the scalp showed angiosarcoma.

The patient was referred for oncological opinion. In view of the extensiveness of his disease, treatment with cytotoxic chemotherapy (vincristine, adriamycin, ifosfamide, mesna) was administered. After four courses of chemotherapy a partial response only was achieved (Figure 2). A few weeks later, the disease reactivated in different areas of the scalp and palliative radiotherapy was given. The patient's condition continued to deteriorate and he became dyspnoeic. A chest X-ray confirmed lung metastases. A few weeks later the patient died of broncho-pneumonia.

Discussion

Angiosarcomas (AS) are extremely rare tumours of vascular origin, constituting less than one per cent of all sarcomas. The preferred areas of origin for AS are the skin and soft tissues, but breast, liver and bone, may also be affected. About 50 per cent of cutaneous AS are found in the head and neck region of the elderly (Kinard *et al.*, 1996). In the majority of cases, there is no clear aetiology but previous radiotherapy and solar exposure have been implicated (Hodgkinson *et al.*, 1979). The most common presenting features are bruise-like macules, dusky red, purple plaques, facial oedema and occasionally extensive ulceration and nodulation (Figure 1). The disease commonly affects the scalp, temple, cheek, but it may also affect the lips, nose, and ears.

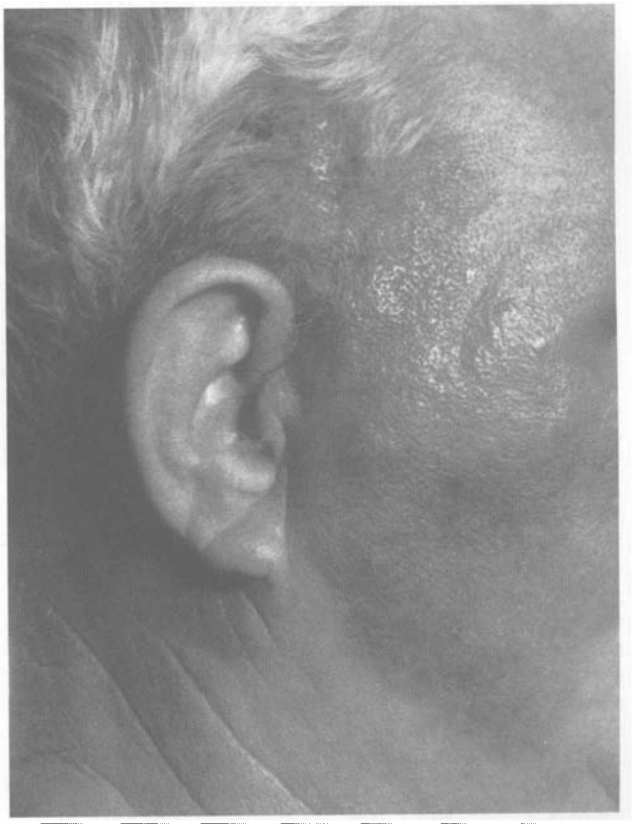

FIG. 1
Extensive angiosarcoma of the scalp and face, showing bruise-like macules and chemosis of eyelids.

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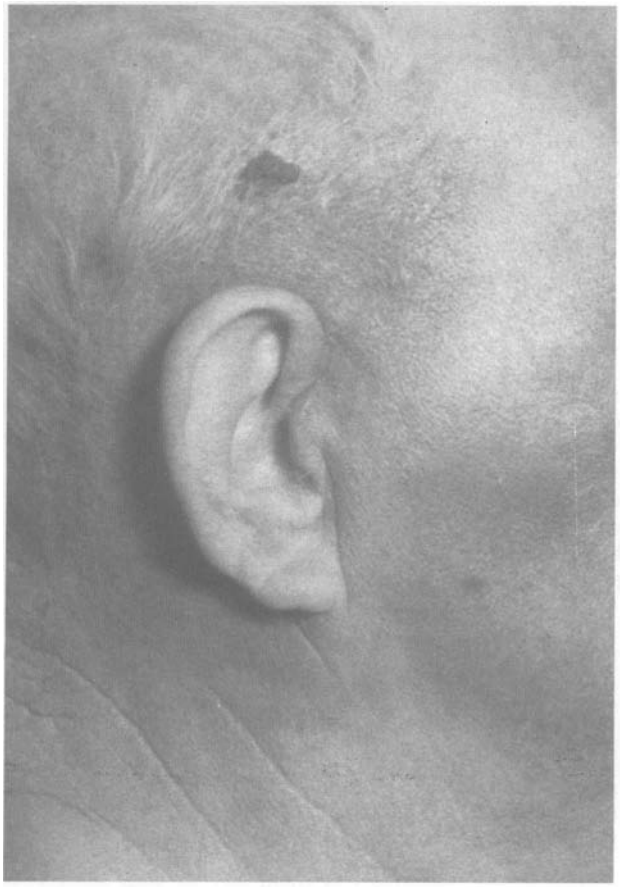

FIG. 2
Partial response of angiosarcoma to chemotherapy.

The histogenesis of AS is controversial, and whether the tumour represents malignant change in blood vessels, or lymphatic vessels, or both, is open to speculation. Electron microscopy has failed, however, to provide a definite answer. However, most authors agree that histological examination, cell marker studies, and absence of Weibel-Polade bodies, support lymphatic derivation (Holden *et al.*, 1987).

Traumatic bruises, infections, and allergic reaction, may be confused clinically with AS. The presence of a preceding history of trauma and other suggestive symptoms of infection or allergy may suffice to make the diagnosis, but biopsy may be necessary. The main challenge in the differential diagnosis, however, comes with other vascular neoplastic diseases, i.e. lymphangiosarcoma, haemangiopericytoma, and Kaposi's sarcoma. Experienced pathologists can usually distinguish these lesions from AS, but close attention to the history, and clinical presentation, can help in differentiation. Lyphangiosarcoma commonly arises on top of lymphoedema, and haemangiopericytoma usually appears as a deep circumscribed mass rather than a cutaneous lesion. Kaposi's sarcoma often has a general distribution, and is usually seen as a late manifestation of acquired immunodeficiency syndrome (AIDS).

The current recommended treatment of AS is radical surgery if complete excision is possible (Maddox and Evans, 1981). Lymph node clearance, in addition to surgical excision, has been advocated for patients with lateralized lesions and palpable lymphadenopathy on presentation (Hodgkinson *et al.*, 1979). Radical surgery alone, however, has been abandoned as a treatment for AS at M.D. Anderson Cancer Centre, because of the very poor long term results (Morrison *et al.*, 1995).

In view of the rarity of AS, the place of radiotherapy in its management is unclear. Radiotherapy, however, has been used as a primary modality and adjuvant to limited surgery with some encouraging results (Rosai *et al.*, 1976; Holden *et al.*, 1987; Morrison *et al.*, 1995). In contrast, Wilson Jones (1964) emphasized the uselessness of radiotherapy in the treatment of AS. This may be due to the frequent delay in diagnosis, and the technical difficulty of administering radiotherapy to the extensive and complex volume at risk. In the last few years, however, a few techniques capable of delivering radiotherapy to the whole head and neck have been described (Sagar and Pujara, 1992; Kinard *et al.*, 1996). It is hoped that through these techniques, an effective means of treating extensive AS can be achieved.

The effectiveness of chemotherapy in AS remains deeply disappointing. In this patient's case, in view of the extent of his disease on presentation, he was treated initially by chemotherapy and his response was only partial (Figure 2).

The overall prognosis of AS is very poor, even when compared with other sarcomas of the head and neck. Whilst the five-year survival rate for AS is less than four to 20 per cent, those with fibrosarcomas, malignant histiocytomas is 60–70 per cent, and those with dermatofibrosarcoma may expect 100 per cent five-year survival rate (Maddox and Evans, 1981; Holden *et al.*, 1987; Wanebo *et al.*, 1992).

The experience with this case is consistent with the experience of others (Holden *et al.*, 1987) that AS are commonly diagnosed late in the course of the disease, which adversely affect the prognosis. But despite the gloomy outlook for AS, there is general agreement that small lesions are amenable to therapy by either surgery, or radiotherapy, or a combination, and that some patients may achieve long-term survival if treated early, hence the importance of early diagnosis.

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