

Case Report

An unusual case of the cutaneous angiosarcoma of the face and scalp

頭面部皮膚血管肉瘤少見病案一例

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Angiosarcoma is an uncommon cutaneous vascular malignancy with a grave prognosis. Classically, it presents with an asymptomatic erythematous to bruise-like macular lesion in the face or scalp. Uncommon presentations include rosacea-like lesions, recurrent facial angio-oedema, cellulitis-like, xanthelasma-like lesions on eyelids causing ptosis and scarring alopecia. Our patient presented with an inflammatory nodular lesion initially masquerading a pyoderma infection. The correct diagnosis was made only after a skin biopsy.

血管肉瘤是一種少見而預後很差的皮膚血管惡性腫瘤。通常表現為頭面部無症狀性紅色或瘀褐色斑片。較少見的表現包括有：玫瑰痤瘡樣疹，復發性面部血管性水腫，蜂窩織炎樣疹，眼瞼黃斑瘤樣疹伴眼瞼下垂，以及疤痕性斑禿。本例表現為疑似感染性的炎性結節狀皮損，及至皮膚活檢始能確診。

Keywords: Angiosarcoma

關鍵詞：頭面部血管肉瘤

Introduction

Angiosarcoma is an uncommon cutaneous vascular malignancy that commonly occurs in elderly patients. It may be classified as idiopathic

angiosarcoma, lymphedematous angiosarcoma and postirradiation angiosarcoma. Presentation includes ulcerative nodules, plaque or bruise-like macule. The following is a report of an elderly patient with angiosarcoma who initially presented as a pyoderma-like lesion.

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Case report

An 84-year-old male presented to us with a forehead lump in November 2002. It started off as a small papule after scratching. It then increased in size quickly within a month and often accompanied by purulent discharge. He was seen in the general out-patient clinic and the lump

shrunk slightly after a course of clavulanate K and amoxicillin trihydrate. His past health was good and there was no history of radiation. On physical examination, a 2.5 x 1.5 cm plaque on right frontal scalp was noted; this was topped with focal crusts and pustules. There was fluctuation on palpation. No cervical lymphadenopathy was detected. The clinical differential diagnoses included kerion, deep fungal infection, atypical mycobacterial infection and squamous cell carcinoma.

Blood tests included complete blood picture, liver and renal functions were normal. Urine for sugar was negative. Wound swab revealed *Pseudomonas aeruginosa*. The lesion was treated initially as an abscess with incision and drainage. A course of ofloxacin was given and the lesion shrunk slightly but then progressed to a fleshy growth (Figure 1).

Incision and drainage was repeated but no more pus could be expressed. In addition, small furuncle-like papules were emerging in the vicinity of the lesion (Figure 2).

An incisional biopsy was subsequently performed which showed lobules of tumour in the dermis (Figure 3). The tumour cells were polygonal with moderately pleomorphic nuclei. Focal formation of irregular dissecting vascular channels lined by atypical, hobnail endothelium and papillary processes was noted (Figure 4). Immunohistochemical studies for endothelial markers, CD31 and CD34 were positive (Figures 5a & 5b) which confirmed the diagnosis of angiosarcoma. The presence of multiple satellite lesions rendered the lesion unresectable. The patient was referred for radiotherapy.



Figure 1. Erythematous and fleshy growth on right frontal scalp with focal crusted areas.



Figure 2. Emergence of satellite nodules in vicinity of the original growth.

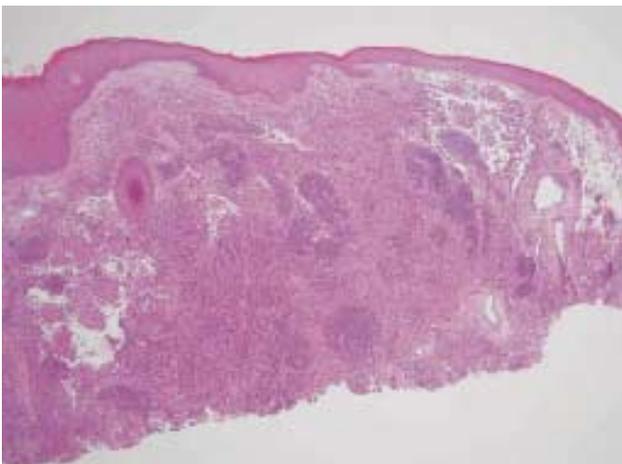


Figure 3. The entire dermis was infiltrated by lobules of tumour cells.

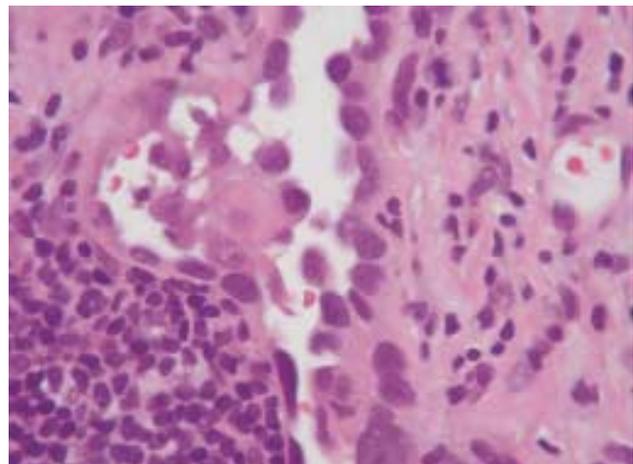


Figure 4. The tumour cells appeared to form irregular dissecting vascular channels in focal areas.

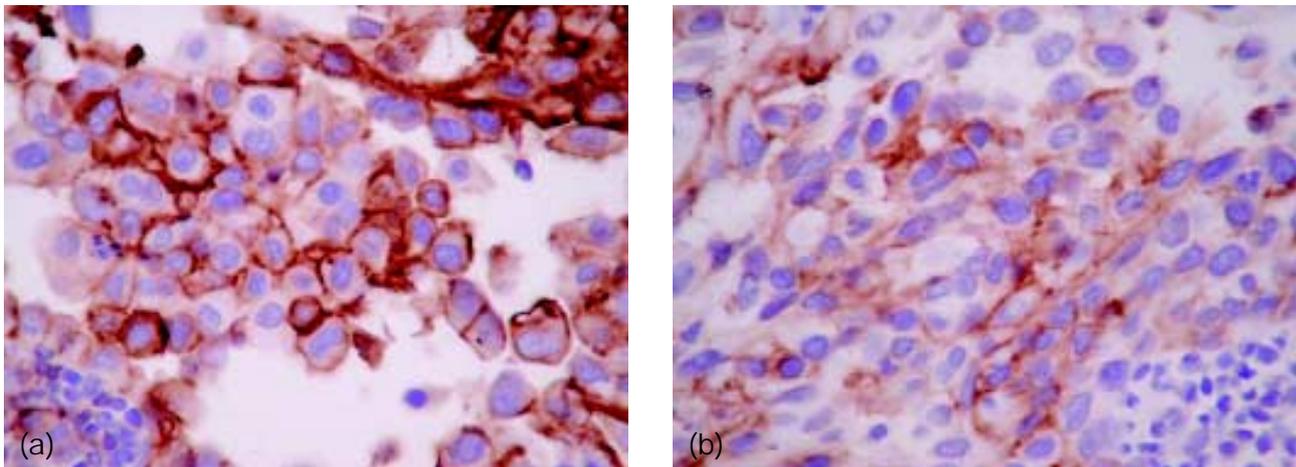


Figure 5. Endothelial markers, CD31 and CD34 were positive.

Discussion

Angiosarcoma accounts for only 2% of all soft tissue tumours. About 60% of all angiosarcomas arose from skin and superficial soft tissue especially in the head and neck region.¹ Cutaneous angiosarcoma (CA) is a rare malignant tumour in which the tumour cells differentiate towards vascular endothelial cells. There are three distinct subgroups: cutaneous angiosarcoma of the face and scalp of elderly patients (Wilson Jones angiosarcoma), cutaneous angiosarcoma associated with lymphoedema and radiation-induced cutaneous angiosarcoma.

CA of the face and scalp of the elderly was first described in 1945, but it was Wilson Jones in 1964 who first distinguished it from other forms of cutaneous angiosarcomas.² This disease affects predominantly elderly with a male to female ratio of 2:1.² No predisposing factor has been identified. It is a highly aggressive tumour and characterised by multifocal infiltrating growth, often extending far wider than the clinical impression. Metastasis occurs after wide local spread in head and neck regions; most commonly to lymph nodes, lungs, liver and bone.³

Clinical presentation of CA can be quite variable. Early lesions may appear as bruise-like areas. The

lesions may be bluish or violaceous; with nodules, plaques, flat infiltrating or ill-defined haemorrhagic areas; there may be haemorrhage or ulceration. Satellite lesions may appear in the vicinity of the main lesion in advanced disease. Atypical presentations such as rosacea-like lesion, recurrent facial angio-oedema, cellulitis-like, xanthelasma-like lesions on eyelids causing ptosis and scarring alopecia had been reported.⁴⁻⁹ In the present case, the initial pyoderma-like presentation was probably due to superimposed infection.

Histologically, CA shows variable degree of differentiation even in the same tumour. The well-differentiated lesions consist of irregular anastomosing vascular channels slanted wide-apart in the dermis and subcutis, which may be misinterpreted as haemangioma or lymphangioma. In poorly-differentiated lesions, there may be solid proliferations of polygonal or spindle-shaped pleomorphic endothelial cells, with poorly formed vascular spaces, mimicking carcinoma and melanoma. In these cases, immunohistochemical studies can be very helpful. Factor VIII-related antigen and *Ulex europaeus* I lectin are positive in most well-differentiated cases. CD34 is more sensitive but less specific. CD31, in contrast, is a highly specific but less sensitive endothelial marker.

The prognosis of aggressive CA is poor, partly due to late presentation or delayed diagnosis. In the largest series reported to date, Holden et al found only 12% of patients survived five years or longer; half died within 15 months of presentation.³ Survival was enhanced if the lesion was less than five cm in diameter and in tumours with a prominent lymphocytic infiltrate. Sex of the patient, location of the lesion, histologic differentiation, or mitotic activity was unrelated to prognosis.¹⁰

The treatment of choice for CA is complete surgical resection. This is often impossible because of the wide extent of lesions and they frequently extend far beyond the clinical margin. For unresectable cases, study showed that wide-field electron beam therapy can prolong survival.¹¹ Other studies also suggested better local control by resection of the macroscopic tumour followed by radiotherapy.^{12,13} Paclitaxel, as a single agent, was found to have substantial activity against angiosarcoma of the face and scalp in a small-scale study done in 1999.¹⁴ Recently, isolated reports on the use of liposomal doxorubicin in angiosarcoma of the scalp showed that it may be a useful alternative to conventional treatment of CA with poor prognosis.¹⁵

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