

Case Report

Cutaneous angiosarcoma: a report of 2 cases

皮膚血管肉瘤：兩宗病例報告

FC Ip 葉方正 and WY Lam 林永賢

A 74 years old Chinese female presented with 9 months history of scalp nodules which gradually increase in number and size with ulceration. Another 73 years old Chinese male presented with 7 months history of scalp nodules which also increase in size with ulceration and bleeding spontaneously. Histopathology from the biopsy of the nodules in both patients confirmed cutaneous angiosarcoma. Grave prognosis is expected in this rare but aggressive neoplasm. Early detection and multidisciplinary approach may improve survival.

74 歲華人女患者近 9 個月來於其頭部出現潰瘍性結節，其數量及大小逐漸增加。另一名患者為 73 歲華人男患者 7 個月來於其頭部出現自行出血性及潰瘍性結節，亦逐漸增大。對此兩名患者的結節進行皮膚活檢，組織學檢查確診為皮膚血管肉瘤。此種腫瘤罕見，生長迅猛而預後很差。早期診斷及多學科綜合治療可提高存活率。

Keywords: Cutaneous angiosarcoma

關鍵詞：皮膚血管肉瘤

Introduction

Angiosarcoma is a high grade malignant neoplasm of endothelial derivation. It can occur in various anatomic sites but has a predilection

for skin and superficial soft tissue, accounting for 60% of cases.¹ About 50% of cutaneous angiosarcoma affects head and neck area in elderly male, particularly the scalp. Clinical presentation in primary cutaneous angiosarcoma varies from early ecchymosis like patch to advance violaceous nodules with haemorrhage and ulceration.

Social Hygiene Service, Department of Health, Hong Kong

FC Ip, MBBS(HK), MRCP(UK)

Department of Clinical Pathology, Tuen Mun Hospital, Hong Kong

WY Lam, FHKCPA, FHKAM(Pathology)

Correspondence to: Dr. FC Ip

Yaumatei Dermatology Clinic, 12/F, Yaumatei Specialist Clinic Extension, 143 Battery Street, Kowloon, Hong Kong

Patient 1

A 74 years old Chinese female presented to us in May 2006 with a 9 months history of enlarging ulcerative nodules in the scalp. She was a housewife and had hypertension on regular

antihypertensive medications. She had no personal or family history of malignancy. She was a non-smoker and non-drinker. The scalp lesions began as small nodules which enlarge and increase in number gradually. There was no preceding trauma, the nodules bled easily.

Physical examination revealed multiple ulcerated violaceous and darkish nodules in vertex and left parietal scalp. The nodules range from 0.5 cm to 2.5 cm in diameter. There was keratosis and oozing in some nodules (Figure 1). There were two 1 cm lymph nodes palpable in left lower cervical region.



Figure 1. Multiple darkish nodules with ulceration, keratosis and oozing in left parietal scalp of patient 1.

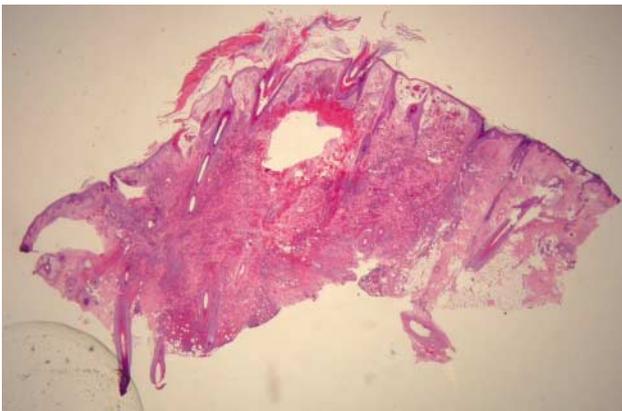


Figure 2. It is lower power view (10X) of the tumour with spindle cell infiltration and proliferation in the dermis, there is also prominent intralesional haemorrhage.

Skin biopsy of the scalp nodule was performed which shown ill-defined spindle cell proliferation in the dermis with prominent intralesional haemorrhage (Figure 2). The neoplasm is composed of plump and oval spindle cells in small fascicles as well as focal capillary and cavernous formation (Figure 3). The cells are plump with active nuclei. There is patchy inflammatory infiltrate in association with spindle cell proliferation. The atypical spindle cells are CD31+ (Figure 4), CD34-, Factor 8-, Human herpesvirus 8-, consistent with angiosarcoma.

Patient 2

A 73 years old Chinese male was seen by us in June 2006 with right scalp nodule for 7 months. The lesion increase in size gradually with episode of spontaneous bleeding. He was otherwise asymptomatic with the lesion. There was no history of trauma prior to onset of the lesion. He has no personal or family history of malignancy. He has history of hypertension and pulmonary tuberculosis more than 30 years ago. He is a non-smoker and non-drinker.

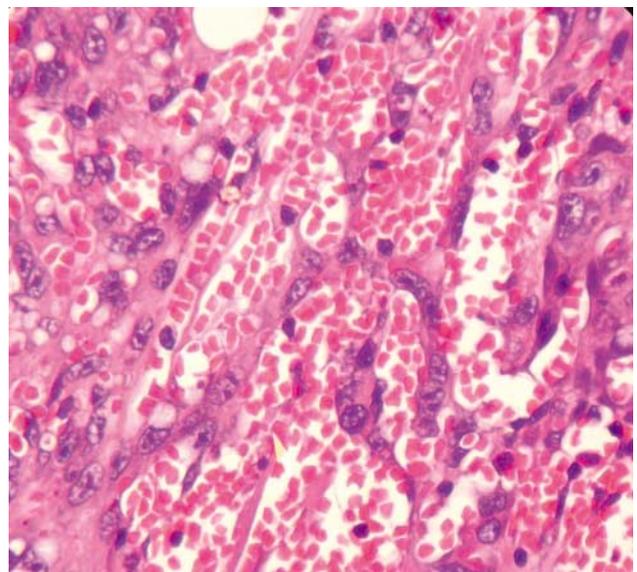


Figure 3. It is high power view (400X) of the abnormal spindle cells in small fascicles and focal capillary and cavernous formation.

Physical examination revealed 2 violaceous nodules in right parietal scalp (Figure 5). The larger nodule measured 1 cm in diameter, it was firm and indurated. The smaller nodule measured 0.5 cm in diameter; it was more fluctuant with scab in the central. There was no cervical lymphadenopathy.

Skin biopsy performed shown a haemorrhagic tumour in the dermis consisting of plump spindle to polygonal tumour cells with moderately

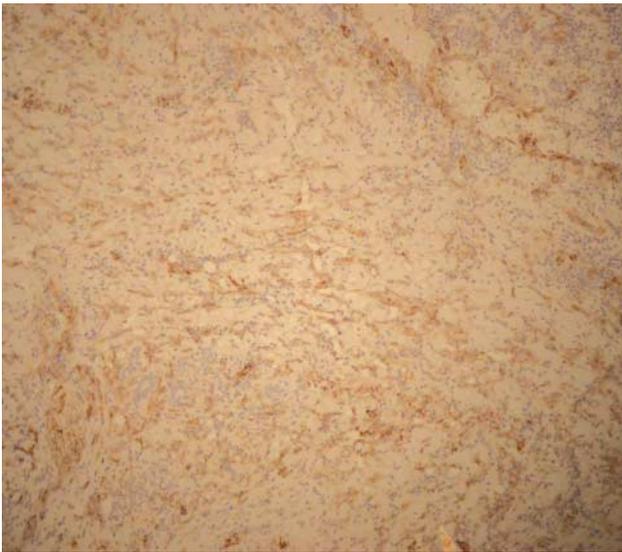


Figure 4. This shown the CD31+ of the abnormal spindle cells.



Figure 5. Violaceous nodule with surrounding diffuse pigmented patch, which indicates local metastasis, in right parietal scalp of patient 2.

pleomorphic nuclei, scattered cytoplasmic vacuoles and intracytoplasmic red cells. The tumour cells show a dissecting growth around the tumour nodule. They form abnormal and irregular vascular channels. These tumour cells are CD31 and CD34 positive. The diagnosis is angiosarcoma.

Discussion

Cutaneous angiosarcoma (CA) was first described in 1945 by Caro and Stubenrauch.² In 1948 Stewart and Treves described association between angiosarcoma and post-mastectomy lymphedema, the Stewart-Treves syndrome.³ Wilson-Jones described cutaneous angiosarcoma primarily affects scalp and face of elderly in 1964.⁴ CA is a rare neoplasm, accounting for <1% of all sarcomas. CA can be classified into primary and secondary. Primary CA was the more common form. In patients with primary CA there had been no previous skin irradiation and lymphedema when compare to patients with secondary CA which usually had either predisposing factor. The most common presentation seen in about half of the cases was an enlarging violaceous nodule over a period of 1 to 3 months, as in patient 2. Other presentations include multiple satellite nodules, as in patient 1; keratotic flesh-toned papule mimic basal cell or squamous cell carcinoma, a discrete vascular tumour such as pyogenic granuloma, or non-alarming ecchymosis and bruise-like patch. Secondary CA is well known to affect area of skin with chronic lymphedema and radiodermatitis. There is a male preponderance in primary CA, male to female ratio of 2-3:1.^{1,5} The average age of onset is more than 74 years. The main clinical differential diagnoses are lymphoma, malignant melanoma, Kaposi's sarcoma, metastatic malignant neoplasm, rarely deep fungal infections such as chromoblastomycosis.

Primary CA is the clonal proliferation of malignant cells express endothelial differentiation. The predominance of head and neck tumour (particularly scalp) raised the speculation of

excessive UV light exposure as the predisposing factor but was inconclusive. The unique vascular density and the anastomotic arrangement of the vessels in the scalp when combined with the effect of UV light might also potentiate oncogenesis. To date, the exact pathogenesis and predisposing factor in primary CA is still uncertain.

CA shows a variable degree of endothelial differentiation. The major histologic type are sinusoidal dissecting, solid epithelioid/spindled and mixed. The sinusoidal dissecting type has irregular proliferation of anastomosing vascular channel lined by abnormal endothelial cells. The solid epithelioid/spindled type shows islands or diffuse sheets of epithelioid/spindled cells without formation of sinusoid or vascular channel. CD31 positivity of the tumour cells is sensitive and specific; it helps to differentiate epithelioid angiosarcoma from carcinoma. Human herpesvirus 8 was not associated with CA as compared to Kaposi's sarcoma.

The two patients presented had the typical age of onset, anatomic site and clinical features of primary CA as described in the literature. There had been no identifiable predisposing factors in both patients. Both had hypertension on regular medication and follow up. They were not immunocompromised and there was no previous radiation or excessive UV light exposure. The prognosis of CA is invariably poor. The overall 5 years survival is 10-35%.¹ Local spread, recurrence and metastasis to regional lymph node and lung are common.⁶ Significant predictors of poor survival include: 1) tumour diameter >5 cm, 2) depth of invasion >3 mm, 3) mitotic figure >3/HPF, 4) positive surgical margin, 5) tumour recurrence and metastases.¹ None of the demographic features have significant association with outcome.

Management is by multidisciplinary approach. Combination of wide local excision, radiotherapy and chemotherapy had been adopted in various institutions. Overall survival had not been improved much with this approach. It is often

difficult to achieve negative surgical margin due to the anatomic site and multiple local metastases on presentation. Combination of surgical excision and post-operative radiotherapy may improve survival especially in younger patients.⁷ Chemotherapeutic agents commonly used are doxorubicin, paclitaxel, cyclophosphamide, vincristine, methotrexate. Disease progression free survival is improved in combination of surgery and chemotherapy with paclitaxel or doxorubicin.⁸ Recent advance of treatment combination with curative radiotherapy and recombinant interleukin-2 administration for CA of scalp shown improve recurrence and metastasis free survival.⁹

Both of our patients underwent wide local excision with post-operative palliative radiotherapy. In view of old age, advanced stage of disease and likely poor tolerability, systemic chemotherapy was not planned for both of the patients.

References

1. Morgan MB, Swann M, Somach S, Eng W, Smoller B. Cutaneous angiosarcoma: a case series with prognostic correlation. *J Am Acad Dermatol* 2004;50:867-74.
2. Caro M, Stubenrauch C. Hemangioendothelioma of the skin. *Arch Dermatol Syph* 1945;51:295-301.
3. Stewart FW, Treves N. Lymphangiosarcoma in post-mastectomy lymphedema. *Cancer* 1948;1:64-81.
4. Wilson-Jones E. Malignant angioendothelioma of the skin. *Br J Dermatol* 1964;76:21-39.
5. Holden CA, Spittle MF, Jones EW. Angiosarcoma of the face and scalp, prognosis and treatment. *Cancer* 1987;59:1046-57.
6. Naka N, Ohsawa M, Tomita Y, Kanno H, Uchida A, Aozasa K. Angiosarcoma in Japan. A review of 99 cases. *Cancer* 1995;75:989-96.
7. Pawlik TM, Paulino AF, McGinn CJ, Baker LH, Cohen DS, Morris JS, et al. Cutaneous angiosarcoma of the scalp: a multidisciplinary approach. *Cancer* 2003;98:1716-26.
8. Fury MG, Antonescu CR, Van Zee KJ, Brennan MF, Maki RG. A 14-year retrospective review of angiosarcoma: clinical characteristics, prognostic factors, and treatment outcomes with surgery and chemotherapy. *Cancer J* 2005;11:241-7.
9. Ohguri T, Imada H, Nomoto S, Yahara K, Hisaoka M, Hashimoto H, et al. Angiosarcoma of the scalp treated with curative radiotherapy plus recombinant interleukin-2 immunotherapy. *Int J Radiat Oncol Biol Phys* 2005;61:1446-53.