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Angiosarcoma presenting as ulcer with hemorrhagic blisters

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Abstract

Angiosarcomas are highly malignant vascular tumors, arising from both vascular and lymphatic endothelium, affecting primarily adult patients. We report a case of a 60 year old lady who presented with hemorrhagic blisters over right preauricular and retroauricular area along with an ulcerated plaque over right temporal scalp of two months duration. Computed Tomography(CT) scan of the head showed soft tissue swelling and no intracranial extension. USG abdomen and CT thorax were normal. Histopathology of the skin lesions showed the upper dermis composed of plump double layered solid, intravascular buds in one area with spindle like morphology and irregular vascular channels lined by atypical endothelial cells, dissecting the collagen. Immunohistochemical staining for CD31, CD34 and VEGF was positive and cytokeratin was negative, confirming the diagnosis of angiosarcoma. Initially we thought of herpes zoster in trigeminal area because of the clinical resemblance. However subsequent investigations pointed towards angiosarcoma. This case report highlights how angiosarcoma may be missed if clinical diagnosis and early investigations are delayed due to mixed presentation and absence of dissemination.

Keywords : CD-31, angiosarcoma, hemorrhagic blisters.

What is known: Cutaneous angiosarcoma is a rare vascular tumor, most commonly presenting as nodules, ulcers, or in rare cases as hemorrhagic blisters, with early dissemination

Introduction

Cutaneous angiosarcoma of scalp, face and neck is a rare malignant vascular tumor, most commonly affecting elderly men with an incidence of approximately 0.01/ 100000. It is more common among Caucasians and it is almost invariably fatal. Presentation as hemorrhagic blisters has been reported in a very few cases. It is well known to cause early dissemination however there are reports showing dissemination occurring in the late stage of disease.

Case report

A 60 year old lady presented with single painful ulcer over right temporal scalp of 2 months duration. The lesion started as hemorrhagic blisters which later ulcerated and increased in size due to repeated scratching. There was no history of fever, loss of weight or appetite, abdominal pain or swelling elsewhere in body. Patient was thin built, otherwise general examination was unremarkable. Cutaneous examination revealed – single well defined ulcerated plaque of size 8 X 6 cm over right temporal scalp, covered with yellowish crust with non-indurated base, friable margins and was freely mobile over underlying structures. There was serosanguinous discharge oozing from the lesion. Multiple hemorrhagic blisters over erythematous, edematous and tender base were present over right cheek, neck & retroauricular region (Figure 1). Aspiration from a blister was bloody and showed red blood cells in roleaux formation and no multinucleated giant cells or atypical cells seen microscopically. CT scan of head showed soft tissue swelling over right temporal scalp with no intracranial extension. CT scan chest and Ultrasonogram of abdomen showed no abnormality. Histopathology from skin lesions showed normal epidermis and vascular channels lined by prominent atypical endothelial cells and the tumor infiltrated the collagen bundles in extensive, disorganised manner(Figure 2 and 3).Immunohistochemistry showed CD31, CD34,VEGF positivity (Figure 4) and was negative for cytokeratin. Patient was referred to Radiation oncology department for further management.

Figure 1 : Ulcerated plaque over right temporal scalp with hemorrhagic blisters over right side of neck

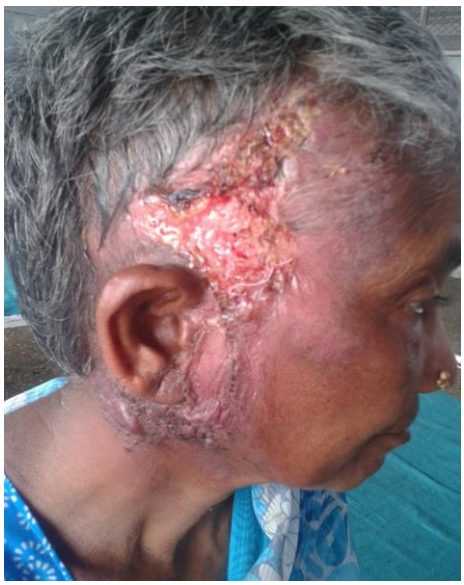


Figure 2 : Histopathology showing extensive dissection of collagen bundles by irregular vascular channels sparing lower dermis and subcutis (H and E) , a. Low power (x100) b.High power (x400)

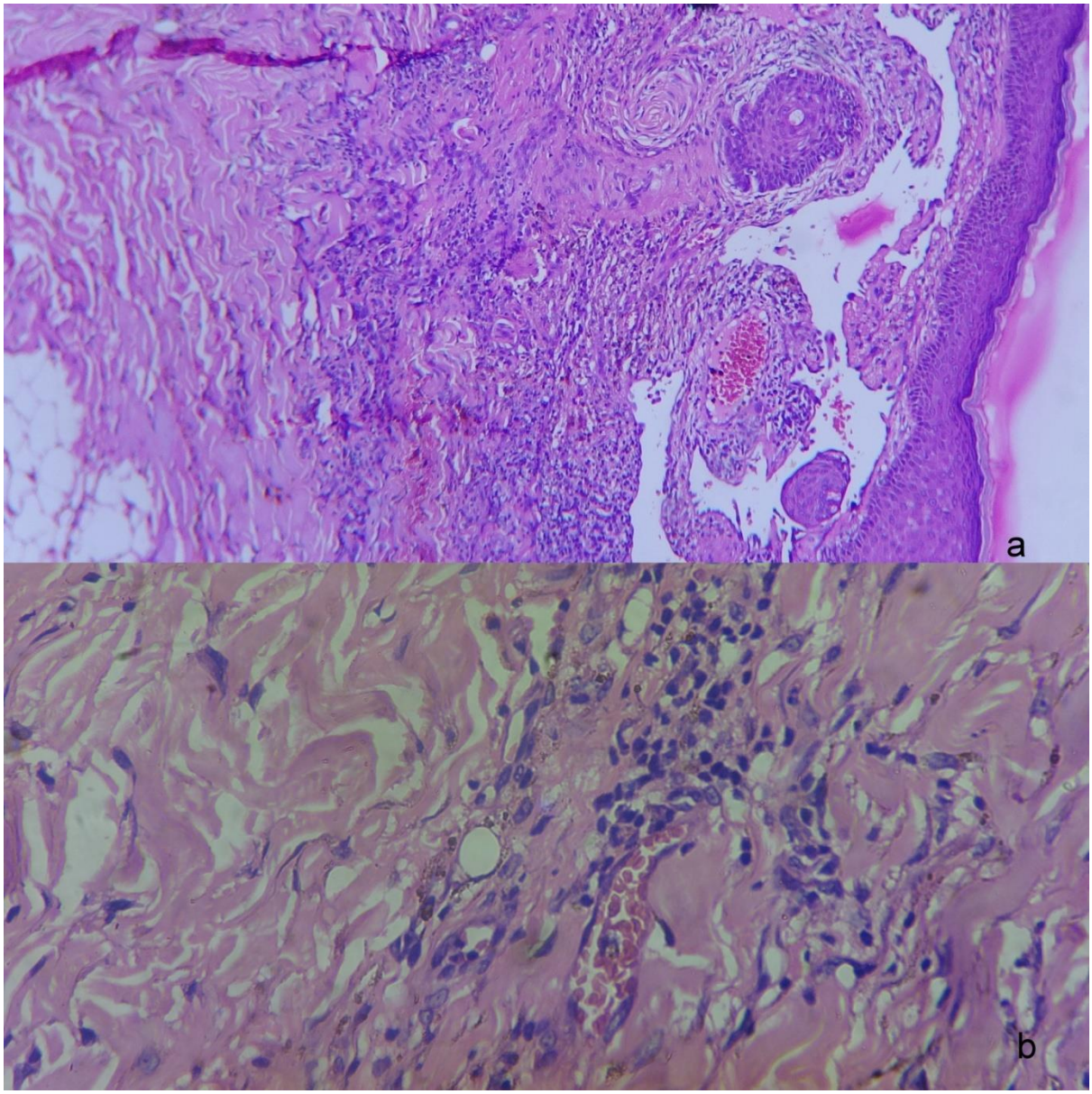
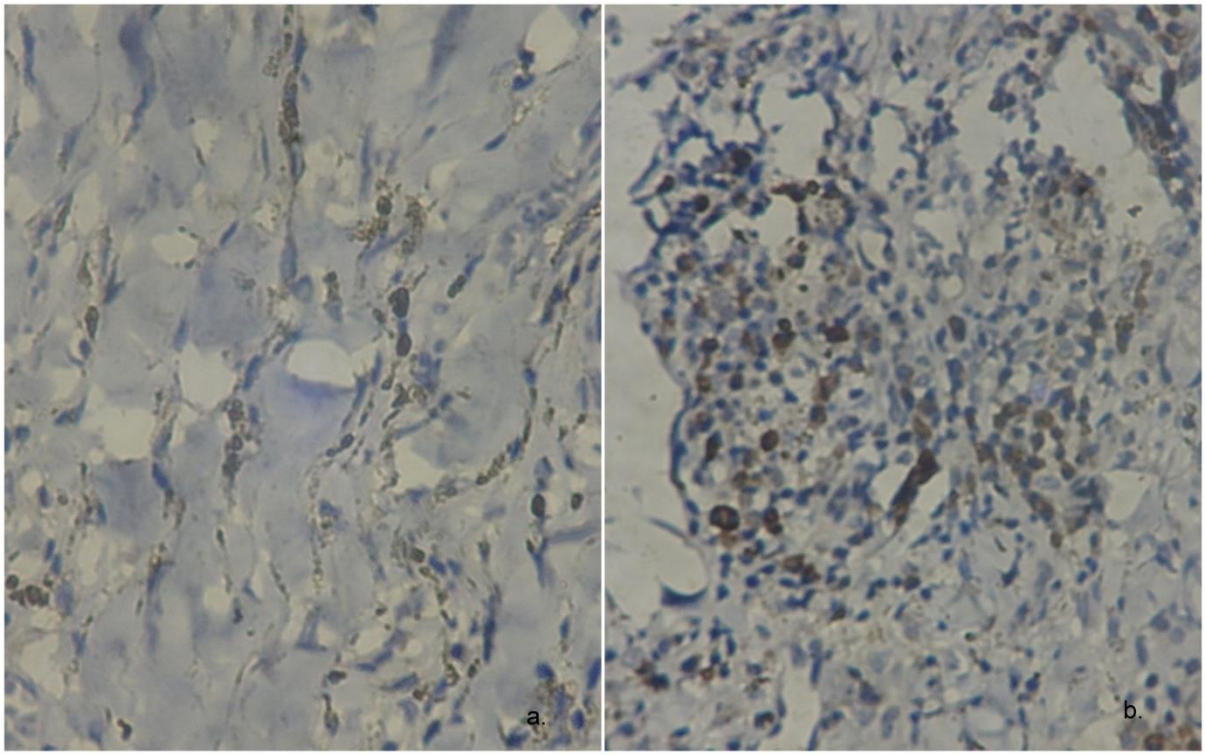


Figure 3 : Immunohistochemistry (x400) positive for, a. CD31 b. CD34



Discussion

Angiosarcomas are highly malignant, uncommon, and aggressive tumor of endothelial or vascular origin[1] with an incidence of approximately 0.01/ 100000 affecting primarily adult patients, especially elderly with male to female ratio of 2: 1. Because of its ambiguous presentation that usually mimics common dermatological conditions, like in our case it mimics herpes zoster, it is often diagnosed late. They account for 1 % of all sarcomas and <1 % of all head and neck cancers. These tumors has predilection for skin and superficial soft tissues in contrast to most other sarcomas that are usually located deep within the body. More than 50% cases are localised to the skin of head & neck regions frequently in upper half of face and scalp.

Cutaneous angiosarcomas almost exclusively occur in three settings: Idiopathic angiosarcoma of face, scalp and neck, also called as Wilson- Jones angiosarcoma; angiosarcoma associated with chronic lymphedema, also known as Stewart -Treves Syndrome ; and post- irradiation angiosarcoma. It may exceptionally occur in vascular malformations,[2] in plexiform neurofibroma,[3] in schwannoma, in xeroderma pigmentosum, in a gouty tophus, chronic vinyl chloride exposure,[4] in association with immunosuppression in organ transplantation etc.
In all types of angiosarcoma, first sign may be an area of bruising, often thought by patient to

be traumatic. Dusky blue or red nodules develop and grow rapidly and fresh discrete nodules appear nearby. In some cases hemorrhagic blisters are a prominent feature, as in our case. As the tumor grows, the edema increases and older lesions may ulcerate. Multifocality is a very frequent finding which makes surgical excision very difficult particularly in face and scalp areas. Dissemination occurs early, with the first visceral deposits usually being in lung and pleural cavity. On histopathology of all three types, vascular channels infiltrate the normal structures in disorganised fashion in a well differentiated tumor, as if trying to line every available tissue with a layer of endothelial tissue. Intense mitotic figures and atypical endothelial cells appear in advanced stages. The collagen is characteristically lined by tumor cells in a pattern that has been described as 'dissection of collagen' which is also demonstrated in early macular lesions of Kaposi sarcoma (HHV-8 positive which is negative in angiosarcoma) and benign lymphangioendothelioma. However these conditions lack mitosis.

Epitheloid angiosarcoma,[5] a histologic variant of angiosarcoma ,clinically resembles angiosarcoma and usually arises in deep soft tissues. It shows solid proliferation of atypical epitheloid cells replacing the dermis and positivity for von Willebrand factor, CD34, Fli-1 and CD31. Cytokeratin positivity is present in up to 50% cases.

Immunohistochemical studies have indicated that antibodies to CD31 are most reliable marker for routine use. Recently antibody against nuclear transcription factor member of the ETS family of DNA binding transcription factors called Fli-1 protein, has been shown to be a fairly specific marker of endothelial cells.
No single treatment modality has proven to be effective. Single most important prognostic factor is the size of lesion, <5 cm tumor size has better prognosis. Because of tumor's propensity for local recurrence and distant metastases, even after prolonged disease free state, long surveillance, especially lung surveillance, is strongly advised.
Our case highlights onset in elderly female, herpes zoster-like morphology and no dissemination to internal organs.

What's new: Mixed presentation with both ulcer and hemorrhagic blisters with no dissemination posing difficulty in early diagnosis of this invariably fatal disease.

References

1. Acharya AS, Sulhyan K R, Ramteke R V, Kunghadkar V Y. Cutaneous lymphangiosarcoma following chronic lymphedema of filarial origin. Indian J Dermatol 2013;58:68-70
2. Rossi S, Fletcher CD. Angiosarcoma arising in hemangioma/vascular malformation: report of four cases and review of the literature. Am J Surg Pathol 2002;26:1319-29.
3. Chaudhuri B, Ronan SG, Manaligod JR. Angiosarcoma arising in a plexiform neurofibroma. Cancer 1980 Aug 1;46(3):605-10.
4. Ghandur-Mnaymneh L, Gonzalez MS. Angiosarcoma of penis with hepatic angiomas in a patient with low vinyl chloride exposure. Cancer 1981;47:1318-24
5. Sundaram M, Vetrichewel TP, Subramanyam S, Subramaniam A. Primary multicentric cutaneous epitheloid angiosarcoma. Indian J dermatol venereal leprol 2011;77:111