Cutaneous Angiosarcoma Associated with the Kasabach–Merritt Syndrome

Dear Editor,

Angiosarcoma is a rare tumour commonly affecting elderly men, usually occurring on the head and neck region.[1] The tumour usually presents as ecchymosis-like plaques, and less commonly with haemorrhage, oedema, ulceration and recurrent facial angioedema.

Kasabach-Merritt syndrome (KMS) may occur in association with a vascular tumour and is characterised by consumption of platelets and coagulation factors within the tumour.[2] This syndrome has been described mostly in children; only a few cases have been associated with angiosarcoma in adults. We report a case of cutaneous angiosaroma of the scalp in an elderly man associated with KMS.

Case Report

An 87-year-old Chinese man, with no prior medical problems, presented with a mass and swelling of the scalp and forehead for 3 weeks. On examination, there were violaceous nodules and plaques on the scalp and forehead with surrounding ecchymoses, and satellite nodules (Fig. 1a).

There were no cervical, axillary or inguinal lymphadenopathy and no hepatosplenomegaly. Histology revealed a tumour in the dermis, composed of sheets of spindle cells with cytoplasmic vacuolation and mitotic activity. The more differentiated parts of the tumour had anastomosing blood vessels lined by malignant endothelial cells (Fig. 1b). On immunohistochemistry, the tumour cells were reactive for CD 31 and CD34, supporting the diagnosis of angiosarcoma.

After biopsy and suturing, there was persistent haemorrhage from the skin biopsy site and he was admitted for observation.

Haematology investigations indicated thrombocytopaenia, with platelets 27,000/μL, haemoglobin 12.0 g/dL and white blood cells (WBC) $8.1x10^3$/μL. Coagulation profile indicated: prothrombin time 10 seconds (range, 9.2 to 11.2), activated partial thromboplastin time 26.7 seconds (range, 27.0 to 36.1), elevated D-dimer 987 μg/L (range, 0 to 324), and low fibrinogen 1.2 g/L (range, 1.61 to 3.99).

This profile suggested KMS, a consumptive coagulopathy - in view of the thrombocytopaenia associated with a raised D-dimer level, and low fibrinogen. Haemorrhage from the biopsy site was controlled with compression packing and platelet transfusion.

Three days later, the patient again had haemorrhage from the biopsy site. Investigations revealed haemoglobin 9.1 g/dL, WBC $6.13x10^3$/μL and platelets 19,000/μL.

Platelet transfusion was done, and intravenous hydrocortisone was commenced, following haematology

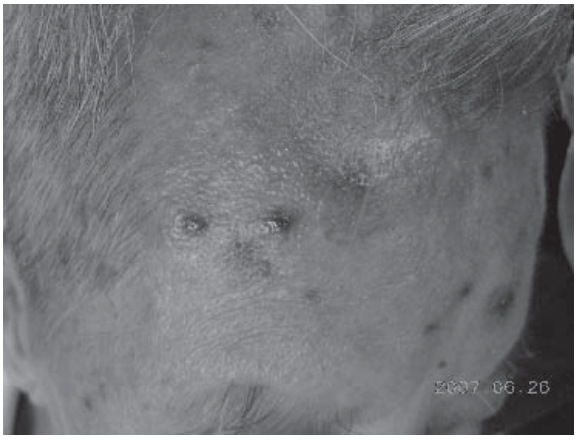

Fig. 1a. Violaceous, erythematous, nodules and plaques over the scalp and forehead.

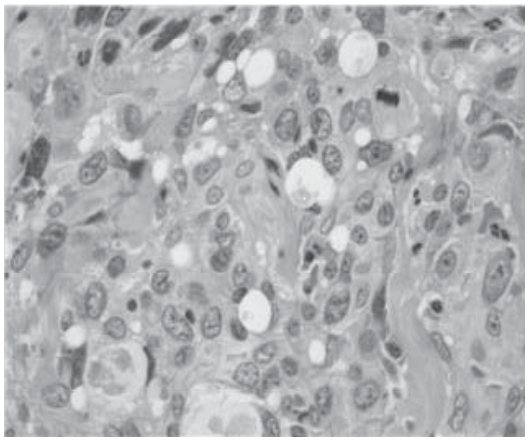

Fig. 1b. Sheets of cells with vesicular and pleomorphic nuclei with mitotic activity. Large cytoplasmic vacuoles are present in some of the cells, destined to fuse and become vascular channels (haematoxylin-eosin stain).

consultation.

Computed tomography (CT) of the brain, chest and abdomen revealed no intracranial haemorrhage or evidence of metastases. Bone marrow examination indicated hypercellular trilineage haematopoiesis with erythroid and megakaryocytic hyperplasia, consistent with a peripheral cause of thrombocytopaenia.

On oncology consultation, weekly paclitaxel was planned followed by radiotherapy to the scalp. Unfortunately, the patient developed congestive cardiac failure with pericardial effusion and chemotherapy was deferred due to the high risk of developing arrhythmia. Radiotherapy was initiated for the angiosaroma. Despite radiotherapy, platelet transfusion and systemic corticosteroids, the patient continued to have thrombocytopaenia with platelet counts ranging from 11,000 to 48,000/μL. He subsequently developed an acute myocardial infarction, leading to death.

Discussion

Cutaneous angiosarcoma is a rare but aggressive tumour that arises from vascular endothelium. Sixty percent of angiosarcomas are cutaneous, and 50% of these occur on the head and neck region. Prognosis is poor with a 5-year survival rate of 12% to 29%.[1]

An unusual presentation of angiosarcoma is the association with KMS. Numerous cases of KMS have been reported in children with tufted angioma and Kaposiform haemangioendothelioma. In contrast, only a few cases of angiosarcoma associated with KMS have been reported in adults. Salameh et al,[3] described a case of metastatic angiosarcoma of the scalp associated with KMS while Moussa et al,[4] reported a case of angiosarcoma of the breast associated with KMS.

The pathophysiology of KMS relates to proliferation of the vascular endothelium within the tumour. Platelets and clotting factors are activated leading to intralesional thrombosis. Continued consumption of platelets and clotting factors initiates fibrinolysis, which results in intralesional haemorrhage and consequent enlargement of the vascular tumour.

During the early stages, the patient can have isolated thrombocytopaenia with a normal clotting screen. A drop in plasma fibrinogen concentration, with a rise in fibrin degradation products (FDPs) and D-dimers marks the onset of coagulopathy.

Haemolytic anaemia is a less frequent mode of presentation, with fragmentation of red cells in the peripheral blood film. One case with metastatic angiosarcoma of the scalp had microangiopathic haemolytic anaemia along with KMS.[3]

Angiosarcoma with KMS is associated with a mortality rate of about 20% to 40%. Fatality in these patients relates to haemorrhage, cardiovascular insufficiency or infection.[4]

Optimal management of angiosarcoma requires complete surgical excision with very wide margins. This is not commonly achieved as the tumour tends to have irregular and diffuse borders that can only be defined histologically.[1] In our patient, surgery was not an option due to the risk of haemorrhage.

Management of KMS aims at halting the proliferation of the vascular tumour, and hence the consumption process. Several modalities have been used, however, no single treatment has been consistently effective. This includes the use of high dose corticosteroids and interferon alpha, which work as anti-angiogenic agents. Other therapeutic modalities include radiotherapy and chemotherapy with vincristine, doxorubicin, paclitaxel and alpha-interferon. Imafuku et al[5] successfully treated a 67-year-old man with early stage angiosarcoma of the scalp complicated by KMS with X-ray irradiation and weekly docetaxel therapy. The authors noted cessation of the consumption coagulopathy and restoration to normal of FDP and fibrinogen levels within 4 weeks.

In summary, KMS associated with angiosarcoma in adults is a rare occurrance. Intractable bleeding may pose a challenge for performing investigations (e.g. skin biopsy, bone marrow examination) and institution of therapeutic options.

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