Clinical Vistas

Ecchymotic groin plaques in an immunocompromised man

A 50-year-old man with a 4-year history of non-Hodgkin's lymphoma presented to the dermatology clinic with painful violaceous plaques in his peri-inguinal area that had slowly enlarged over 3 weeks. A year before presentation the patient had undergone allogeneic stem-cell transplantation and was considered in remission except for mild, chronic hepatic graft-versus-host disease, for which he received several immunosuppressive medications, including prednisone, tacrolimus and mycophenolate mophetil.

Physical examination revealed tender ecchymotic indurated plaques over the anterior portion of the patient's thighs and suprapubic area (Fig. 1). The patient was otherwise well. A wedge biopsy of one of the plaques revealed a poorly circumscribed hemorrhagic malignant tumour filling the dermis that consisted of sheets and lobules of poorly differentiated epithelioid and focally spindled malignant cells with frequent mitotic figures. On histologic examination, the tumour cells demonstrated strong cytoplasmic staining for the endothelial cell marker CD31 and strong nuclear staining for the oncogene Fli-1, and no staining for lymphoid and epithelial markers. These findings were consistent with a diagnosis of angiosarcoma. Whole-body CT and positron emission tomography scans were negative for nodal and metastatic spread. Liposomal doxorubicin therapy was started and the dose of the patient's immunosuppressive regimen reduced, which led to resolution of his cutaneous lesions.

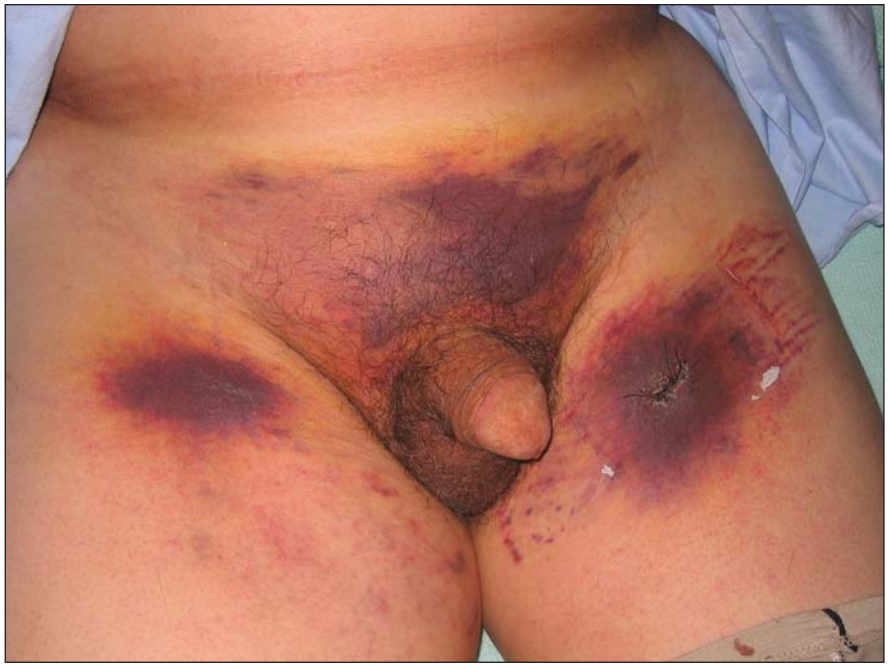

Fig 1: Ecchymotic indurated plaques over the anterior portion of the thighs and suprapubic area in an immunocomprised patient.

Angiosarcoma is a rare malignant tumour of endothelial origin, accounting for 2% of all sarcomas. Unlike most sarcomas, it has a predilection for skin and subcutaneous tissues. About 50% of cases occur on the face and scalp, predominantly in elderly men.[1] Angiosarcoma can arise in the setting of chronic lymphedema, as in Stewart–Treves syndrome following mastectomy and lymph-node dissection for breast cancer. Cutaneous angiosarcoma can also occur after radiation treatment. Immunosuppression is a likely risk factor for angiosarcoma, as it is for other malignant diseases.

The clinical presentation of angiosarcoma varies. It typically appears as a slowly expanding erythematous plaque; a deceptively benign-appearing ecchymotic or bruise-like presentation may be observed, which may delay diagnosis.[1] Histologically, malignant endothelial cells in angiosarcoma are often poorly differentiated and may show positivity with various endothelial markers, including Factor VIII, *Ulex europaeus*-1 lectin, Fli-1, CD34 and CD31. Human herpesvirus 8 DNA is usually absent in cases of angiosarcoma, in contrast to Kaposi's sarcoma. The clinical differential diagnosis for angiosarcoma is broad. In our patient it included neoplastic processes (lymphomatous skin infiltrates and Kaposi's sarcoma), infectious conditions (ecthyma gangrenosum and bacillary angiomatosis), vasculitis and coagulopathies (lupus anticoagulant syndrome).

Angiosarcoma is an aggressive tumour that tends to recur locally and to metastasize to lymph nodes, lungs, liver, brain and bone. The prognosis is poor, with a reported 5-year-survival rate of 10%–35%.[1,2] Patients with lesions less than 10 cm in size have significantly better survival rates,[2] which emphasizes the importance of early diagnosis. Depending on the extent of the disease, treatment modalities include surgical excision, radiotherapy, and chemotherapy with paclitaxel or liposomal doxorubicin.[3]

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Competing interests: None declared.

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DOI:10.1503/cmaj.051586