Cutaneous Angiosarcoma Mimicking Panniculitis in the Lower Extremities

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Sir,

The 36-year-old, otherwise healthy male admitted to our clinic with a complaint of a red swelling on his right ankle for 3 months. A history of lymphedema with an unknown etiology in the lower extremities was present in the patient. On the physical examination; a painful, red-livid erythematous, soft plaque; which was surrounded by a yellowish ecchymotic color was observed on the medial malleolus of the right ankle [Figure 1]. A punch biopsy was performed with prediagnosis of traumatic panniculitis and erythema nodosum. In the histopathologic examination; eosinophilic stained, atypical epithelioid cells with prominent nucleoli trying to form several vessel structures were observed [Figure 2]. In the immunohistochemical study, tumoral cells stained strongly positive with CD31 and friend leukemia integration 1 (FLI-1) [Figure 3]. Ki-67 proliferation index was very high (60%). A diagnosis of cutaneous angiosarcoma was made with clinical and histopathological findings. At the follow-up, 3 weeks after the first admittance, a rapid growth in the diameter of the lesion and a pronounced vascular character was observed [Figure 4]. No distant metastases were detected in the positron emission tomography-computed tomography examination. A wide local surgical excision was performed which was followed by 33 sessions of local radiotherapy postoperatively. After 16 months, in his regular follow-ups, metastatic disease involvement in the lungs was detected, and chemotherapy was planned.

Angiosarcoma is a rare and malignant soft tissue tumor with a high mortality and aggressive course, which develops from endothelial cells lining blood vessel walls. Cutaneous angiosarcomas constitute one-third of all angiosarcomas. Classical cutaneous angiosarcoma usually presents on the head and neck region of elderly male patients.[1] Other than the classical form, lymphedema-associated angiosarcomas and postradiation angiosarcomas are well described. Chronic lymphedema-related angiosarcoma, which was identified for the first time in 1948 by Stewart and Treves, constitutes 10% of all cases.[1] Although it most commonly settles in the upper extremities following mastectomy and lymph node dissection, rare cases of...
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Angiosarcoma of the lower extremity on a background of lymphedema have also been reported.[2,3]

Clinical presentation of cutaneous angiosarcoma might be variable. It can manifest as slightly erythema, contusion-like bruises, red-purple papulonodules, and necrotic plaques. Differential diagnosis includes Kaposi sarcoma, pyogenic granuloma, amelanotic melanoma, cutaneous lymphoma, and cutaneous metastasis.

Histopathology shows atypical ectatic vascular spaces lined by pleomorphic endothelial cells with high mitotic activity. Immunohistochemical studies with endothelial markers such as CD31, CD34, Faktor VIII, Podoplanin (D2-40), and FLI1 are expected to be positive, the latter being the one with highest specificity.[4]

Cutaneous angiosarcoma has one of the worst prognoses among malignant cutaneous tumors, and a 5-year life expectancy has been reported as 10%–30%.[5] Early diagnosis is important to improve patient survival. This case is reported to remind the possibility of angiosarcoma when purple-red, painful lesions, or nonhealing ulcers develop on lymphedematous extremities.

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Conflicts of interest
There are no conflicts of interest.

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