Cutaneous Angiosarcoma

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Abstract
Cutaneous angiosarcoma is a rare malignant tumor showing blood or lymphatic vessel differentiation, corresponding to <2% of all sarcomas. It preferably affects elderly, with predilection for the head and neck. Diagnosis is frequently late due to the early interpretation by the patient as a benign lesion similar to ecchymosis, which explains its aggressiveness with high metastasis and recurrence rates. We report the case of an elderly man whose histopathologic diagnosis confirmed the clinical suspicion of cutaneous angiosarcoma.

Introduction
Angiosarcoma represents <2% of soft tissue sarcomas and 5% of all malignant skin tumors. It is a rare and aggressive neoplasia showing blood or lymphatic vessel differentiation that can occur in any anatomical location, although the head and neck are the most common sites [1]. These tumors are characterized by difficult surgical margin delimitation, high proneness for local recurrence, and distal metastases. Horizontal dissemination can result in the involvement of extensive skin portions, with the tumor progressing usually along vascular neck structures towards the mediastinum. Prognosis is poor with a 5-year survival rate...
ranging from 12 to 34%. Aiming at a clinical demonstration of this rare tumor, we report a case of location in the forehead, emphasizing its aggressiveness and the relevance of an early diagnosis [1, 2].

Case Report

An 86-year-old man noticed an erythematous-violaceous macule on his forehead that evolved to a painless extensive and friable tumor within 5 months. He denied previous trauma or exposure to chemical agents. Dermatologic examination showed a violaceous tumoral lesion of approximately 15 cm on his forehead with a central ulceration area (Fig. 1). Histopathologic examination revealed irregular anastomosing vascular channels in the dermis, lined by endothelial cells with enlarged and hyperchromic nuclei, dissecting collagen bundles, and surrounding vascular structures and eccrine ducts (Fig. 2). Immunohistochemical study was positive for CD31 (Fig. 3) and negative for CD34 and HHV-8. Skull computed tomography (CT) revealed an extensive and heterogeneous lesion affecting the dermis and subcutaneous and sparing the skullcap of the frontotemporal region, the upper eyelids, and the glabella. Additionally, the patient presented mediastinal lymphadenomegaly at thorax CT scan. Brain magnetic resonance angiography, transthoracic echocardiography, and abdominal CT scan did not show significant alterations. Based on these results, the diagnosis of cutaneous angiosarcoma was made, and palliative chemotherapy and antihemorrhagic radiodermotherapy were initiated.

Discussion

Angiosarcoma can be classified into three clinical variants: cutaneous angiosarcoma associated to chronic lymphedema (also called lymphangiosarcoma), radiation-induced angiosarcoma, and primary cutaneous or idiopathic angiosarcoma. Angiosarcoma originated by chronic lymphedema, also known as Stewart-Treves syndrome, occurs predominantly in the upper limbs of women submitted to radical mastectomy with axillar emptying, but is also observed in patients with congenital, traumatic, or infectious lymphedema. Postradiation angiosarcoma is also seen with greater frequency in women, on average after 10 years of radiotherapy for treatment of breast carcinoma, and is nowadays the second most frequent subtype. Cases of primary cutaneous angiosarcoma, originated in absence of previous irradiation or lymphedema, usually affect the scalp or frontal region of elderly men, as in the reported case [1, 3, 4].

Treatment should be customized in accordance with disease staging, but due to the rarity of its presentation, there are no adequate randomized prospective studies for therapeutic standardization. Surgical resection with ample margins is the treatment of choice and the only potential cure if free margins are reached. In most cases however, complete excision is not possible due to the extension and multifocal nature of the disease [2, 5].

In the present case, palliative chemotherapy with paclitaxel and antihemorrhagic radiotherapy was chosen based on the stage of the patient, classified as IV because of the distal lymph node involvement. The tumor was considered inoperable due to its size and location. The patient received three chemotherapy cycles with paclitaxel 80 mg/m² intravenously on days 1, 8, and 15, and radiotherapy with 30 Gy in 10 fractions in the first scheme and 20 Gy in 5 fractions in the second scheme. Despite an initial favorable response, the patient evolved
with new lesions and intermittent bleeding. Unfortunately he died from undetermined cause at home 7 months after the start of therapy.

Lymph nodes and the lung are the most frequent sites of metastases, and in case of lymph node involvement the prognosis is similar to that of distant metastasis. Due to the difficulty of resection with free margins, tumors located in the head and neck or those exceeding 5 cm (2 inches) have the worst prognosis, hence early diagnosis is fundamental to increase the survival rate for such patients [2, 3, 6].

**Statement of Ethics**

All ethical determinations were followed.

**Disclosure Statement**

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Fig. 1. Purplish tumorous lesion on the frontal region with central ulceration.
Fig. 2. Irregular anastomosing channels in the dermis, paved by endothelial cells with enlarged and hyperchromatic nuclei, surrounding vascular structures and eccrine ducts (HE, ×400).
Fig. 3. Positivity for CD31 (CD31, ×400).