IJD Case Report

An unusual manifestation of cutaneous angiosarcoma: a case report of a rare tumor

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Cutaneous angiosarcoma is a rare tumor of the head and neck region, most commonly affecting the elderly male. Its presentation varies from a small plaque to multifocal nodules. Differentiating this tumor from other conditions, such as hemangiomas, Kaposi sarcoma, squamous cell carcinoma, and rosacea, is sometimes difficult. Herein, we present a case of a 73-year-old male with a small oozing lesion on the scalp for more than two months. He had a history of scalp irradiation for tinea capitis in his childhood. Also, he experienced multiple basal cell carcinomas on his scalp a few years ago. Skin biopsy revealed infiltrations of malignant neoplastic lesions composed of proliferated pleomorphic tumoral cells with hyperchromatic nuclei and some epithelioid features arranged as sheets and irregularly shaped vascular spaces mostly devoid of red blood cells. Neoplastic cells were diffuse and strongly positive for D2-40, CD31, CD34, and Ki67 but negative for C-myc and CK. Cutaneous angiosarcoma should be considered in the differential diagnoses of scalp lesions, particularly in older men.

Keywords: cutaneous angiosarcoma, scalp, radiation, CD31+, tumor

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INTRODUCTION

Cutaneous angiosarcoma is one of the rare soft tissue tumors; only 10% of soft tissue sarcoma cases of the head and neck region are related to cutaneous angiosarcoma [1]. Its presentation differs from a bluish or violaceous nodule (single or multiple) to plaques and flat infiltrating areas with occasional bleeding or ulceration [2-4]. Angiosarcoma is known for its great mimicker properties; it can be confused with a variety of diseases, such as Kaposi sarcoma [5], sebaceous cysts [6], ecchymosis [7], and squamous cell carcinoma [1]. Herein, we report a very unusual presentation of angiosarcoma on the scalp of an elderly man with an old history of radiation exposure.

CASE PRESENTATION

A 73-year-old man presented to the outpatient dermatology office with mildly painful oozing on the right side of his scalp. Since two months ago, he had noticed a mild swelling of spontaneous onset with oozing on the right side of his scalp, which slowly spread into adjacent tissue. Oozing continued despite several courses of systemic antibiotics (Figure 1).

On examination, there was small ulceration with clear oozing on a background of ill-defined yellowish discoloration on the scalp. He had a history of scalp irradiation for tinea capitis in his childhood. Also, he experienced multiple basal cell carcinomas on his scalp before. No lymphadenopathy was detected

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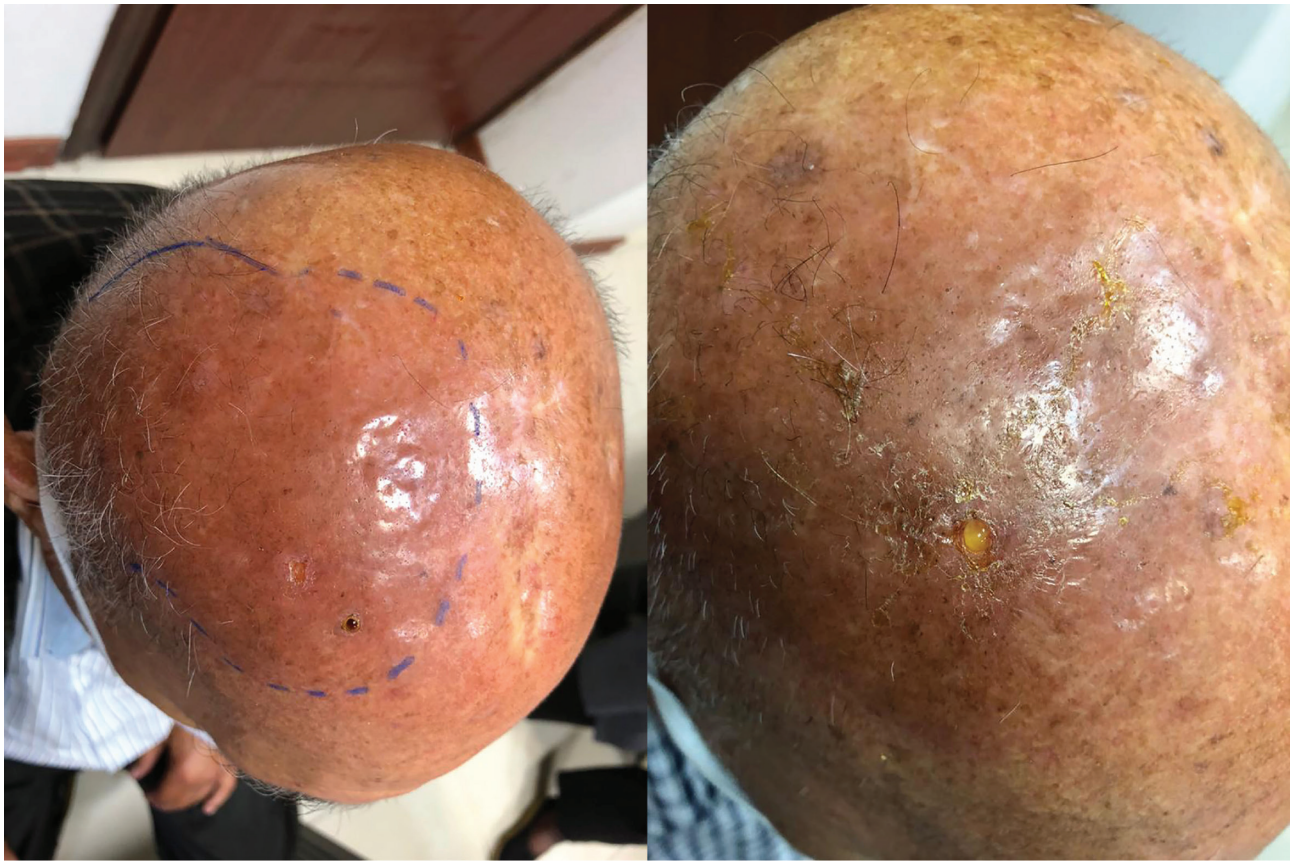

Figure 1. Painful oozing lesion on the scalp of a 73-year-old man with prior irradiation history.

on examination. He was otherwise a healthy man.

Skin biopsy from the nodular lesion of the scalp revealed infiltrations of malignant neoplastic lesions composed of proliferated pleomorphic tumoral cells with hyperchromatic nuclei and some epithelioid features arranged as sheets and irregularly shaped vascular spaces, mostly devoid of red blood cells (Figure 2). Neoplastic cells were diffuse and strongly positive for D2-40, CD31, CD34, and Ki67 but negative for C-myc and CK. The tumor deeply extended to subcutaneous fat with foci of perineurial and perimuscular invasion (Figure 3). We found no necrosis or infiltration into the lymphatics or blood vessels. Hence, cutaneous angiosarcoma, at least grade II, was concluded. Other investigations, such as the chest X-ray and contrast-enhanced computed tomography of the head and neck, reported normal findings. A metastatic workup revealed no systemic metastasis. We referred the patient to the oncology department.

Ethical consideration

The patient provided informed consent for the publication of this case report.

DISCUSSION

Angiosarcoma is a malignant neoplasm of endothelial cells lining the blood vessels. They can also involve the viscera such as the breast, spleen, and liver [8]. Scalp angiosarcoma usually happens in old men in the late decades of their life. The estimated male-to-female ratio is 3:1 [2]. Although most lesions are not associated with a pre-existing condition [9], age, irradiation, chronic lymphedema, trauma [10], and immunosuppression in renal transplant patients [11] are considered the predisposing factors for angiosarcoma. According to its wide variety of probable etiologies, at present, some variants of cutaneous angiosarcoma, including radiation-induced, lymphedema-induced (Stewart-Treves syndrome), primary breast angiosarcoma, sporadic cutaneous age-related angiosarcoma, and soft tissue angiosarcoma have been recognized [3,11].

Cutaneous angiosarcomas are well known for their great mimicry characteristics and delayed

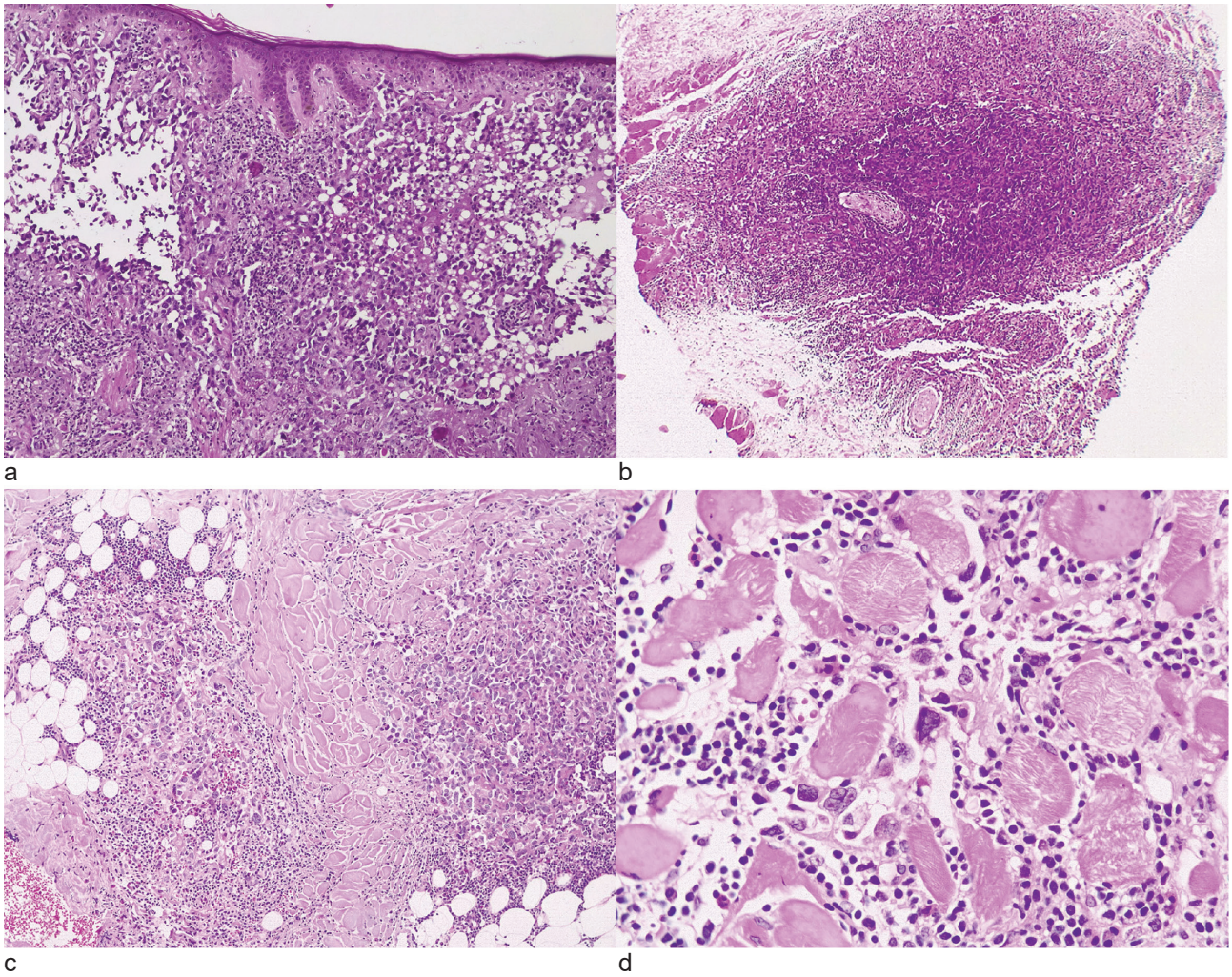

Figure 2. (a) Infiltrations of malignant neoplastic lesions composed of proliferated pleomorphic tumoral cells with hyperchromatic nuclei and some epithelioid features arranged as sheets and irregular vascular spaces mostly devoid of red blood cells (H&E, 100×). (b) Perineural involvement of malignant cells (H&E, 40×). (c, d) Muscular involvement of malignant cells (H&E, c:100×, d:400×).

diagnosis. Its presentation differs from a bluish or violaceous nodule (single or multiple) to plaques and flat infiltrating areas with occasional bleeding or ulceration [2-4]. Our case was unique in presenting with just a discharging ulcer on the scalp. To our knowledge, this rare presentation had not been reported before.

Angiosarcomas often widely invade the dermis. The poorly differentiated variants involve deep structures such as fascia and subcutis. Angiosarcoma can occur around vascular channels, where sheets of cells and undifferentiated morphologic cellular features can be seen. Angiosarcomas exhibit varying degrees of differentiation. In well-differentiated types, some of the normal vascular endothelium's morphologic and functional features are preserved—so-called low-grade angiosarcomas. Sheets of poorly differentiated pleomorphic cells that look like a carcinoma with the possible presence of hemorrhage, distorted architecture, and large cells with hyperchromatic, pleomorphic nuclei are considered high-grade tumors [2,11]. Both types can be associated with extensive local growth. However, the grade of the tumor does not have any correlation with survival [2].

Immunohistochemical markers include von-Willebrand factor, CD34, CD31, VEGF, Ulex europaeus agglutinin 1, and factor VIII antigen [3]. CD31 is the most sensitive and specific marker for endothelial differentiation, while the von Willebrand factor and Ulex europaeus agglutinin are less important [8]. In addition to CD31+ and CD34+ cells, our case was positive for D2-40 and Ki-67.

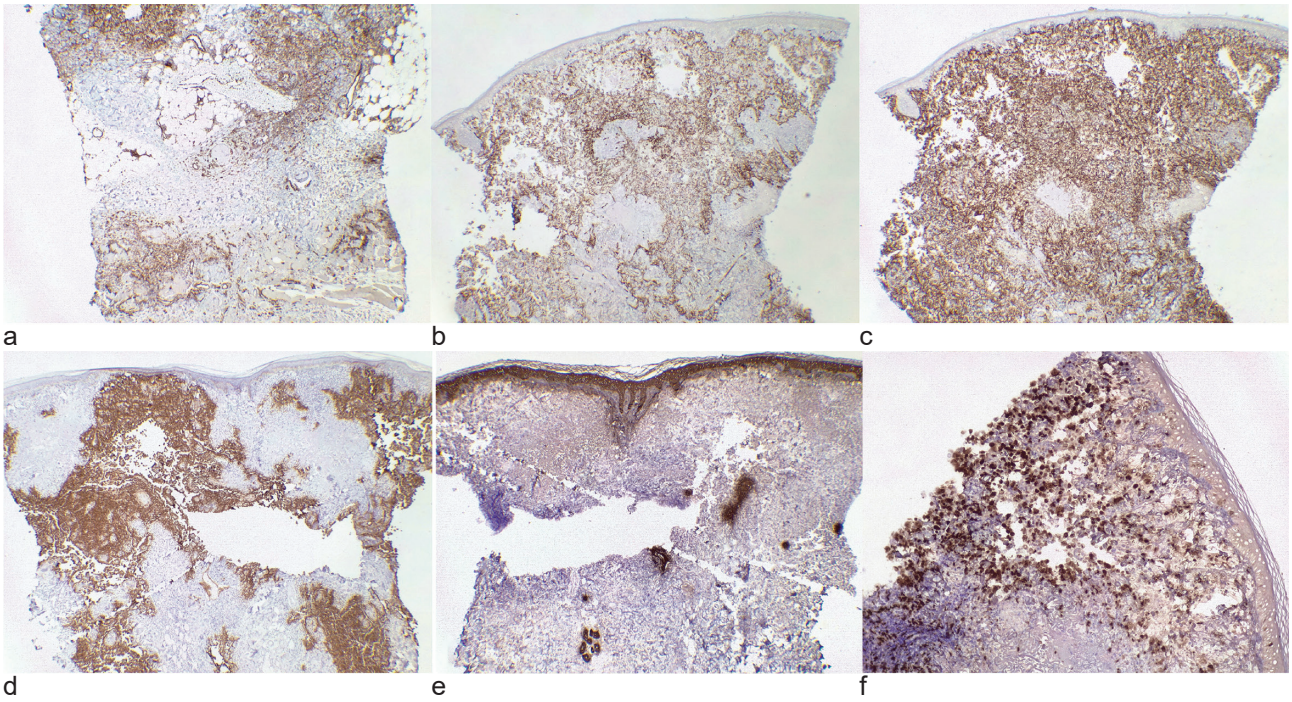

Figure 3. Immunohistochemistry staining of scalp lesion biopsy. (a, c) Positive for CD34, (b) Positive for CD31, (d) Positive for D2-40, (e) CK staining for ruling out an adenocarcinoma, (f) Ki67 positive for 20-25% (40×).

Angiosarcoma aggressively invades local and distant areas via lymphatic and hematogenous vessels. Angiosarcoma has a high rate of regional nodal involvement (20–30%), and the lungs are the most common site of metastasis. The liver, spleen, and bone are after that. The characteristic findings of lung metastasis are cystic pulmonary lesions with thin walls and pneumothorax. Our case had no regional or distant metastasis. Generally, the prognosis of head and neck angiosarcoma is miserable, with a five-year survival rate of just 10–20% [2,8].

Treatment is usually by radical excision and reconstruction. Radiotherapy and chemotherapy are used for those with distant metastasis or unresectable tumors [2,11].

CONCLUSION

The great mimicry properties of angiosarcoma, besides its different rare presentations and the poor prognosis of the metastatic stages of the tumor, warns of precise suspicion of this rare tumor, especially for the lesions of the scalp and face in old men.

Authors contributions

The patient was managed by Dr. Zargari and Dr. Azimi, who participated in data acquisition. Dr. Mesbah performed pathologic analysis and interpretation. Dr. Zargari and Dr. Azimi drafted the manuscript. All authors revised the manuscript critically for important intellectual content and approved the final version.

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