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Case Report

A Novel Presentation of Cutaneous Angiosarcoma: A Case Report and Review

Abstract

We report a case of a 70-year-old male, with slowly widening induration, ulceration, and oozing for 3 months on the scalp and face. The diagnosis of aggressive cutaneous angiosarcoma was made on histopathology and immunochemistry from the biopsy material from the involved area of the skin.

Keywords: *CD31+, CD34+, cutaneous angiosarcoma, mimicking cellulitis, vimentin*

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Introduction

Cutaneous angiosarcoma is a rare soft tissue tumor found more often in the head and neck region. Less than 5% cases of soft tissue sarcomas involve head and neck, 10% of which are angiosarcomas.[1] They may present as single or multiple bluish or violaceous nodules or plaques or flat infiltrating areas which occasionally bleed or ulcerate.[2] They are known to be great mimickers and may mimic rosacea, local bruise, eczema, Kaposi sarcoma, and scarring alopecia to elucidate a few. This leads to wider extension and delay in diagnosis. We report a case of cutaneous angiosarcoma with unsuspecting presentation and rarity.

Case Report

A 70-year-old male presented with progressively extending, mildly painful, ulcerated, and red indurated skin lesion involving the left side of the scalp for 3 months and the face for 10 days. Three months back, he noticed a painless swelling with oozing on the left side of the vertex of the scalp. This slowly spread into surrounding tissue. Some of the area ulcerated and black eschar formed over it. The mild ooze continued and the surrounding peripheral skin hardened and progressed to involve the left side of the forehead, the periorbital region, and the upper left cheek. Besides, he had been unable to open his left eye for 10 days [Figure 1].

On examination, the left side of the scalp and face had an erythematous ill-defined to well-defined, indurated, stony hard, nontender plaque with mild scaling involving root of the nose, upper cheek, upper and lower eyelids, side of the forehead from midline to pinna, mastoid, frontal, temporal, parietal, and vertex regions. There was decreased density of hair in the left eyebrow with areas of prominent follicular openings. The parietal and temporal regions were covered with black eschar and areas of oozing and crusting up to the upper left mid-occipital region [Figure 2]. The patient could not open his left eye due to the indurated upper eyelid. The submental, submandibular, anterior cervical, posterior cervical, and supraclavicular lymph nodes of both sides were 2–3 cm in diameter, discrete, mobile, firm, and nontender.

His blood glucose was 145 mg/dl fasting and 280 mg/dl postprandial. Hemogram, renal and liver function tests were within normal limits. A provisional diagnosis of cellulitis with diabetes mellitus was kept and he was treated accordingly. The discharge sent for the culture revealed *Acinetobacter*. The treatment was changed as per the sensitivity profile. After initial improvement on antibiotics, the erythema and induration not only persisted but also progressed.

Fine-needle aspiration cytology from the left posterior cervical lymph nodes and left cheek revealed round to spindle-shaped cells, arranged in clusters with anisocytosis and coarse chromatin. They were probably malignant cells of mesenchymal origin along with acute inflammatory cells and necrosis.

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Skin biopsy from the edge of the plaque in the scalp divulged ulcerated epidermis and a tumor in the dermis. The tumor cells were pleomorphic, and the nuclei were hyperchromatic, plump, and spindle-shaped. The cells with occasional mitosis were arranged in sheets and at places lined with irregular slit-like spaces, infiltrating into the surrounding collagenous stroma and arrector pili muscle [Figure 3]. No infiltration into the lymphatics or blood vessels could be detected. Immunohistochemistry done in the biopsied skin showed positive CD34, CD31, vascular endothelial growth factor (VEGF), and vimentin while pan cytokeratin was negative [Figure 4]. It was thus confirmed to be cutaneous angiosarcoma. X-ray chest was normal. X-ray and contrast-enhanced computed tomography (CECT) head and neck revealed no evidence of skull invasion or intracranial extension. Ultrasound abdomen and CECT chest and abdomen showed no systemic metastasis. The patient was referred to oncology department but succumbed in 6 months.

Discussion

Angiosarcomas are rare malignant tumors derived from endothelial cells that line the blood vessels. They constitute only 1%–2% of all soft tissue sarcomas while sarcomas themselves account for <1% of all malignancies.[3] They can be either cutaneous or visceral involving the breast, spleen, and liver.[3] Scalp angiosarcoma usually occurs in elderly men in the seventh to eighth decade of life with an estimated male-to-female ratio of 3:1.[2] However, 14 cases (including the present case) reported from India have a male-to-female ratio of 1.8:1 and an early age of onset [Table 1].

Predisposing factors for angiosarcoma include trauma, chronic lymphedema, irradiation, and age. Due to its wide range of plausible etiologies, it has been categorized into lymphedema-associated, radiation-induced, primary breast angiosarcoma, sporadic cutaneous angiosarcoma (age related), commonly located on the head and neck region,[13] and angiosarcoma of the soft tissue.[17] Although chronic lymphedema is the most widely recognized predisposing factor in angiosarcoma of the skin and soft tissue, other common predisposing factors may be trauma, irradiation, and age *per se*.[17] Our case had a short clinical

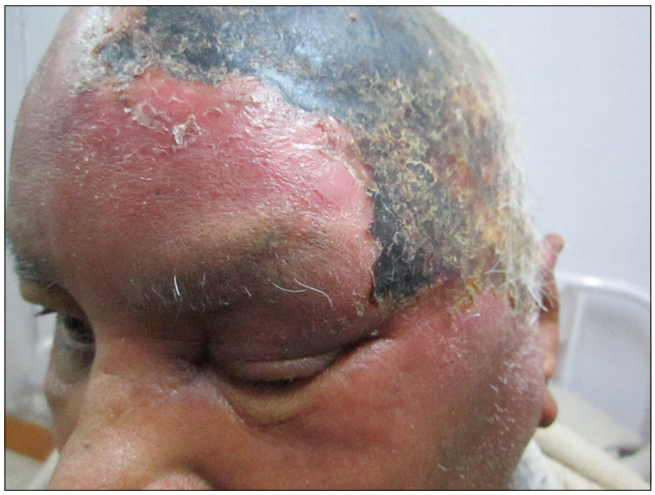

Figure 1: Frontal view of an erythematous, indurated, stony hard plaque with central eschar on the scalp associated with edema of the left upper eyelid

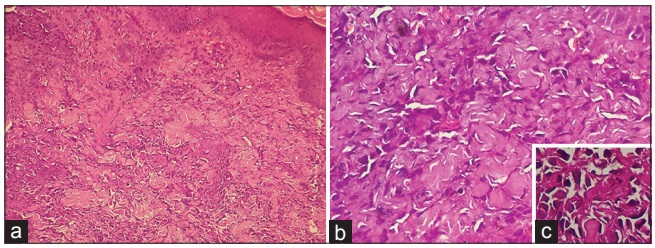

Figure 3: (a) Histopathology slide showing skin biopsy specimen with tumor cells (H and E, ×100). (b and c) Pleomorphic, hyperchromatic, plump tumor cells with spindle-shaped nuclei in the dermis. Cells with occasional mitosis were arranged in sheets and at places lined with irregular slit-like spaces, infiltrating into surrounding collagenous stroma and arrector pili muscle (inset) (×400)

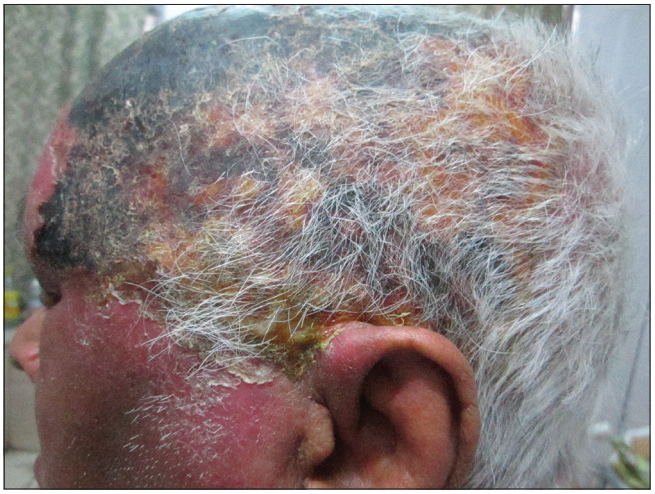

Figure 2: Lateral view of the same patient showing involvement of most of the left side of scalp, cheek, and pinna

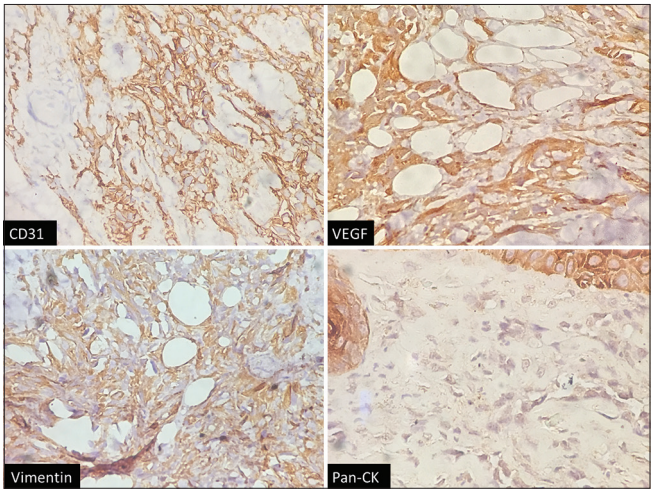

Figure 4: Immunohistochemical stains show positive CD31, vascular endothelial growth factor, and vimentin and negative for pan cytokeratin

Table 1: List of Indian cases published till date with their age, site, presentation, and prognosis

Serial number	Author	Age (years) and sex of patient	Site of involvement	Clinical presentation
1	Arora *et al*. 2008[4]	40/male	Lateral aspect of the right eyebrow	Malignant melanoma-like lesion
2	Sundaram *et al*. 2010[5]	80/female	Temporal, parieto-occipital scalp	Cellulitis-like lesions with metastasis to liver
3	Bhagwat *et al*. 2005[6]	47/female	Scalp	Chronic nonhealing ulcer with metastasis to cervical lymph nodes, liver, bone (hip joint), and lung
4	Kharkar *et al*. 2012[7]	66/female	Tip of the nose	Erythematous ulcerated plaque
5	Ambujam *et al*. 2013[8]	65/male	Both eyelids, forehead, and parietal region of the scalp	Indurated plaques mimicking Hematoma around both eyes
6	Banswal and Patil 2014[9]	43/female	Scalp	Large ulcerative nodular growth with spread to underlying cranium
7	Gupta *et al*. 2009[10]	55/female	Scalp	Infected scalp granuloma
8	Sharma *et al*. 2012[11]	27/male with XP	Scalp	Chronic nonhealing ulcer with spread to superior sagittal sinus
9	Siddaraju *et al*. 2008[12]	20/male	scalp and forehead	Multiple nodules with erosion of underlying bone
10	Bhardwaj *et al*. 2005[13]	65/male	Scalp	Ulcernodular growth
11	Dhanasekar *et al*. 2012[14]	60/male	Scalp	Multiple nodules mimicking SCC
12	Rajinikanth *et al*. 2007[15]	70/male	Scalp	vascular reddish-colored macules and blue-black nodules with ulcerated surface
13	Deshpande *et al*. 2012[16]	74/male	Scalp	Nonhealing ulcer
14	Present case	70/male	Scalp	Erythematous, indurated, nontender plaque with areas of eschar and cervical lymphadenopathy

SCC – Squamous cell carcinoma, XP - Xeroderma pigmentosum

history of 3 months of stony hard progressively increasing plaque in the scalp with cervical lymphadenopathy. This only stands to reason that these findings suggest that lymphedema had occurred secondary to the infiltrating tumor mass rather than the tumor secondarily developing in lymphedema, as has been described earlier. The lymphedema in this case could either be due to compression or blockage of the lymphatics by the invading tumor mass.

Cutaneous angiosarcomas are known to be great mimickers and may present as single or multiple, bluish or violaceous nodules, plaques, or flat infiltrating lesions.[2,17] Occasionally, they may ulcerate or bleed. They have often being described in literature to mimic rosacea,[18] local bruise,[8] eczema,[17] Kaposi sarcoma,[19] scarring alopecia,[20] sebaceous cysts,[21] and rhinophyma[22] and thus delaying the diagnosis [Table 2].

Histologically, angiosarcomas often extensively involve the dermis, and the poorly differentiated tumors invade deep structures, such as fascia and subcutis. The histologic patterns in which it can occur are around vascular channels, sheets of cells, and cells with undifferentiated morphologic features. Low-grade angiosarcomas are well-differentiated lesions that retain some of the functional and morphologic properties of normal vascular endothelium. In high-grade tumors, cells are poorly differentiated and sheets of pleomorphic cells may resemble a carcinoma. They may have areas of hemorrhage, distorted architecture, and large cells with hyperchromatic, pleomorphic nuclei.[2,10]

Table 2: List of conditions and clinical presentations cutaneous angiosarcoma has been reported to mimic

Serial number	Author	Disease mimicking
1	Trinh *et al*.[17]	Eczema
2	Mentzel *et al*.[18]	Rosacea
3	Ambujam *et al*. 2013[8]	Hematoma
4	Shehan and Ahmed[19]	Kaposi sarcoma
5	Knight *et al*.[20]	Scarring alopecia
6	Pan *et al*.[21]	Sebaceous cyst
7	Lo Presti *et al*.[22]	Rhinophyma
8	Gupta *et al*.[10]	Infected scalp granuloma
9	Arora *et al*.[4]	Malignant melanoma
10	Goldenberg[23]	Pyogenic granuloma
11	Dhanasekar *et al*.[14]	SCC, hemangiomas
12	Brightman *et al*.[24]	Malignant melanoma
13	Riahi and Cohen[25]	Keratoacanthoma

SCC – Squamous cell carcinoma

Immunohistochemical markers include von Willebrand factor, CD34, CD31, Ulex europaeus agglutinin 1, VEGF, and factor VIII antigen.[17] The most sensitive and specific marker for endothelial differentiation is CD31. Von Willebrand factor is fairly specific, and Ulex europaeus agglutinin has poor sensitivity and specificity, thus were not done.[3] In our case, besides, CD31+ cells and CD34+ cells, VEGF, and vimentin were also positive.

Angiosarcoma has a tendency for aggressive local as well as distant metastasis through lymphatic and hematogenous

routes. The rate of regional nodal involvement is said to be 20%–30%, which is higher than most other sarcomas. The most common site for metastasis is lungs. Thin-walled, cystic pulmonary lesions and pneumothorax are characteristic findings.[2,3] Other less common sites of distant spread are liver, spleen, and bone. This case had metastasis to the lymph nodes while metastasis to other organs could not be detected. The overall prognosis for patients with angiosarcoma of the head and neck remains dismal, with a reported 5-year survival rate of approximately 10%–20%.[2,3] The delay in diagnosis due to initial misinterpretation, advanced age at presentation, lesions >5 cm in greatest dimension, multifocal nature of lesions at presentation, poor histological grade, lesions that initially cannot be surgically resected, and lesions with greater depth of invasion are the major factors responsible for poor prognosis reported in cutaneous angiosarcomas.[3,10]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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