

Spot the Diagnosis

Erythematous plaque with ulceration on the scalp

Arunachalam Narayanan¹, Kalimuthu Abinaya¹, Pampa Ch Toi², Devinder Mohan Thappa¹

¹Department of Dermatology and STD, ²Department of Pathology, Jawaharlal Institute of Postgraduate Medical Education and Research, Gorimedu, Puducherry, India.



***Corresponding author:**
Arunachalam Narayanan,
Department of Dermatology
and STD, Jawaharlal Institute
of Postgraduate Medical
Education and Research,
Gorimedu, Puducherry, India.
narayanan359@gmail.com

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A 58-year-old male farmer presented to our clinic with a red plaque with ulceration and oozing for 4 months. The patient noticed a red lesion over the scalp after a local trauma, which gradually progressed to result in an erythematous, indurated plaque of size 10 × 8 cm over the frontoparietal scalp [Figure 1] during the next 4 months. There were multiple smaller, skin-

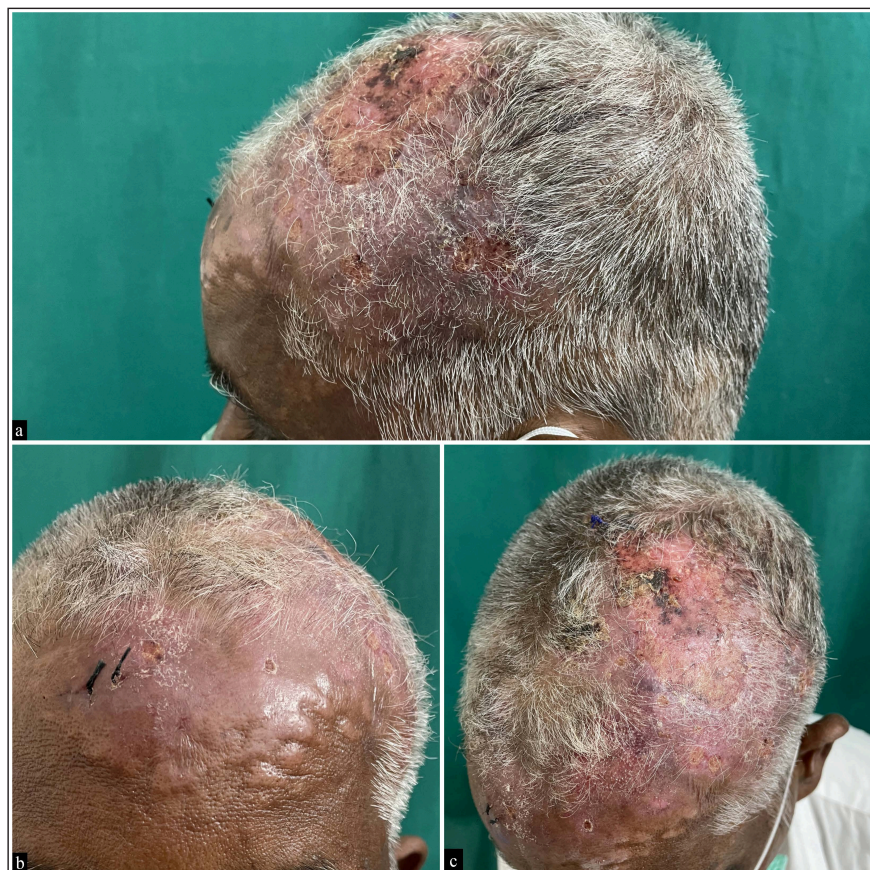


Figure 1a-c: Erythematous, indurated plaque of size 10 × 8 cm with central ulceration over the frontoparietal scalp, multiple smaller skin-colored plaques along the border.

colored satellite papules, and plaques along the border. There was no lymphadenopathy. He visited a general practitioner who made a diagnosis of scalp psoriasis and treated him with apremilast and tar shampoo unsuccessfully. The patient visited our tertiary care center after developing ulcers and oozing over the lesion. There were no systemic symptoms or loss of weight. He had no headache or vision problems. Histopathology of a biopsy specimen taken from the scalp revealed numerous irregularly-shaped anastomosing vascular channels with atypical endothelial cells [Figure 2]. These atypical endothelial cells had an epithelioid morphology

with vesicular nuclei and prominent nucleoli. These cells infiltrated the deeper tissues. There was an increase in mitotic activity (Ki-67 proliferative index-70%). There were numerous ectatic blood vessels surrounded by tumor cells. Few lymphocytic infiltrates were seen interspersed between these tumor cells. However, no necrosis/hyaline globules were identified. On immunohistochemistry, the tumor cells were positive for vimentin, CD31, and CD34, and negative for pan-CK.

What is your diagnosis?

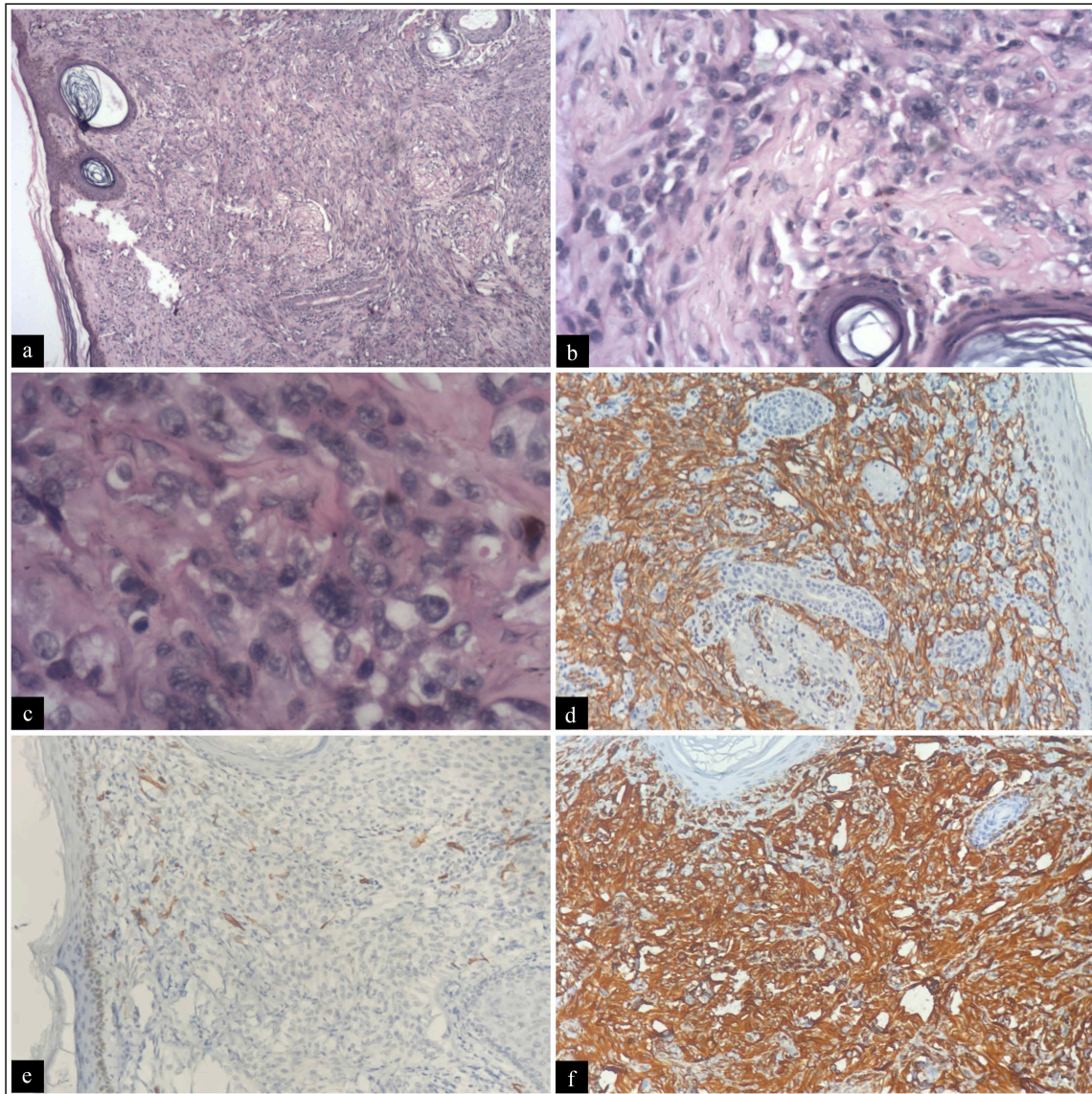


Figure 2a-f: (a) Histopathology revealed numerous irregularly-shaped anastomosing vascular channels with atypical endothelial cells infiltrating the deeper tissues. (H&E, $\times 4$), (b) There were numerous ectatic blood vessels surrounded by tumor cells. (H&E, $\times 20$), (c) The atypical endothelial cells had an epithelioid morphology with vesicular nuclei and prominent nucleoli, and increased mitotic activity. (H&E, $\times 40$), few lymphocytic infiltrates were seen interspersed between these tumor cells. However, no necrosis/hyaline globules were identified, (d) Immunohistochemistry revealed positive for CD31, (e) CD34, and (f) vimentin.

Angiosarcoma

DISCUSSION

Angiosarcomas are a clonal proliferation of malignant cells with endothelial differentiation. They occur commonly in Caucasian men and individuals >70 years of age.^[1] Angiosarcoma can be primary, post-lymphedema, or postradiotherapy.^[2] Exposure to vinyl chloride, thorium dioxide, arsenic, radium, and anabolic steroids have been linked to the development of angiosarcoma. The most common form of angiosarcoma is cutaneous angiosarcoma in the elderly. More than 50% of lesions involve the head and neck area.

The lesions typically start appearing as a benign-appearing bruise-like patch on the central face, forehead, or scalp. Later on, patients start developing facial edema, and with time, lesions progress into violaceous nodules that bleed easily and ulcerate. The lesions spread centrifugally, covering large areas of the head and neck. On the other hand, pediatric cutaneous angiosarcomas are small, unifocal, and occur in the lower extremities in girls.

Angiosarcomas that occur following chronic lymphedema appear as firm, coalescing violaceous nodules or plaques against a background of non-pitting edema.^[3] More than 90% of such cases occur following mastectomy and lymph node dissection (Stewart-Treves syndrome). In such cases, the most common site is the inner aspect of the upper arm. Usually, lymphedema occurs 4–27 years before the onset of angiosarcoma.

Angiosarcoma can also occur following radiotherapy.^[4] The greater incidence of breast-sparing surgery has resulted in post-irradiation angiosarcoma arising within breast parenchyma and overlying skin. They present as infiltrated plaques or nodules over the irradiated area. The average time for the development of angiosarcoma after radiation is 6 years.

Histopathology of angiosarcoma demonstrates well-differentiated areas of the anastomosing network of sinusoidal vessels lined by a single layer of endothelial cells with nuclear atypia. These cells infiltrate collagen bundles and adipose tissue splitting them apart. The cells have more nuclear pleomorphism and mitotic activity in less well-differentiated areas. They form papillary projections. Poorly differentiated regions don't have luminal formation. Angiosarcomas stain positively for CD31, CD34, ERG, c-MYC, and Fli-1.^[5]

Patients have a poor prognosis with <15% survival over a 5-year period. Patients with angiosarcoma require surgical excision with wide margins. The recurrence rate and metastasis risk are high. Other treatment options include paclitaxel chemotherapy, propranolol plus vinblastine-based metronomic therapy, thalidomide, and radiotherapy.^[6] Early recognition of this condition is essential for prompt diagnosis and appropriate treatment.

Declaration of patient consent

Patient's consent is not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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