An elderly man with an asymptomatic plaque on the scalp

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CLINICAL PRESENTATION

A 75-year-old male presented with a history of an asymptomatic swelling over the parietal region of the scalp for one year with a rapid increase in size over the past 3 months. On examination, a solitary 4 × 4 cm well-defined erythematous to bluish plaque with ulceration and central scarring was noted (Figure 1). The lesion was non-tender, firm, and bled on palpation. There was no associated regional lymphadenopathy. He did not have any other cutaneous lesion elsewhere in the body. Systemic examination was normal. An elliptical incisional skin biopsy was performed and sent for histopathological examination and immunohistochemistry.

what is your diagnosis?

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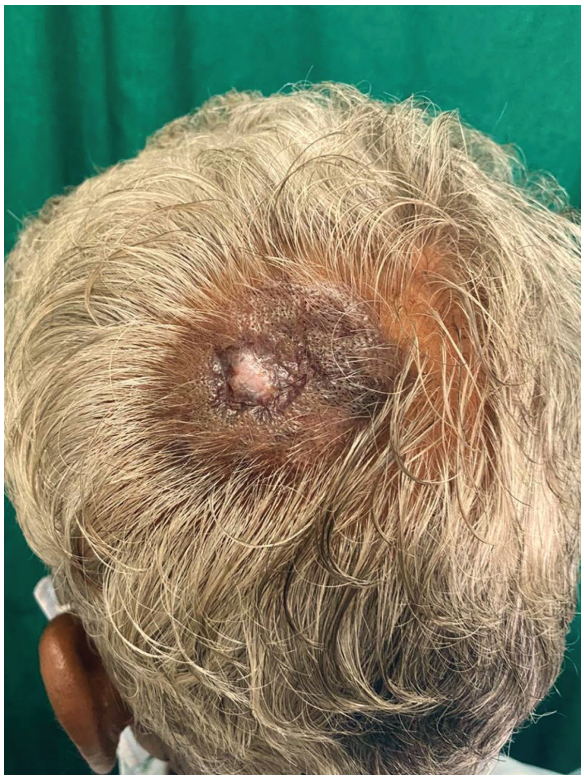

Figure 1. A solitary 4×4 cm well-defined erythematous to bluish plaque with ulceration and central scarring.

DIAGNOSIS

Angiosarcoma

Microscopic findings

Histopathological examination showed epidermis with hyperkeratosis, acanthosis, and follicular plugging. The dermis had scattered vascular spaces lined by plump endothelial cells with intraluminal papillations and nuclear atypia. The vascular channels dissected the collagen bundles of the dermis. There were scattered chronic inflammatory infiltrate and areas of hemorrhage (Figure 2). Immunohistochemically, the specimen was positive for markers CD31 and CD34 (Figure 3).

DISCUSSION

Angiosarcomas are aggressive malignant endothelial-cell tumors of vascular or lymphatic origin [1]. They are common among the elderly, with no gender predilection [2]. They can arise from any soft-tissue structure or viscera. In the skin, the scalp is the most common site involved, followed by the neck, breast, extremities, and trunk [1]. Angiosarcomas are further classified into cutaneous, lymphedema-associated, post-radiation, and soft tissue angiosarcomas [1]. Risk factors include radiation, exogenous toxins like arsenic and vinyl chloride, and familial syndromes like neurofibromatosis and Maffucci syndrome [1,6].

Cutaneous angiosarcomas typically present as

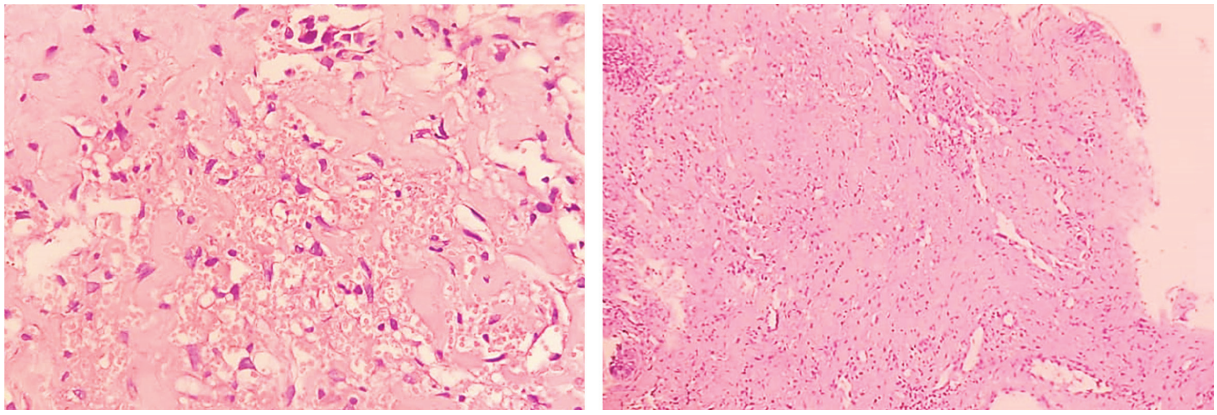

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Figure 2. (a) Endothelial cells exhibiting features of nuclear atypia; (b) Vascular channels traversing the collagen bundles (H & E, 10×).

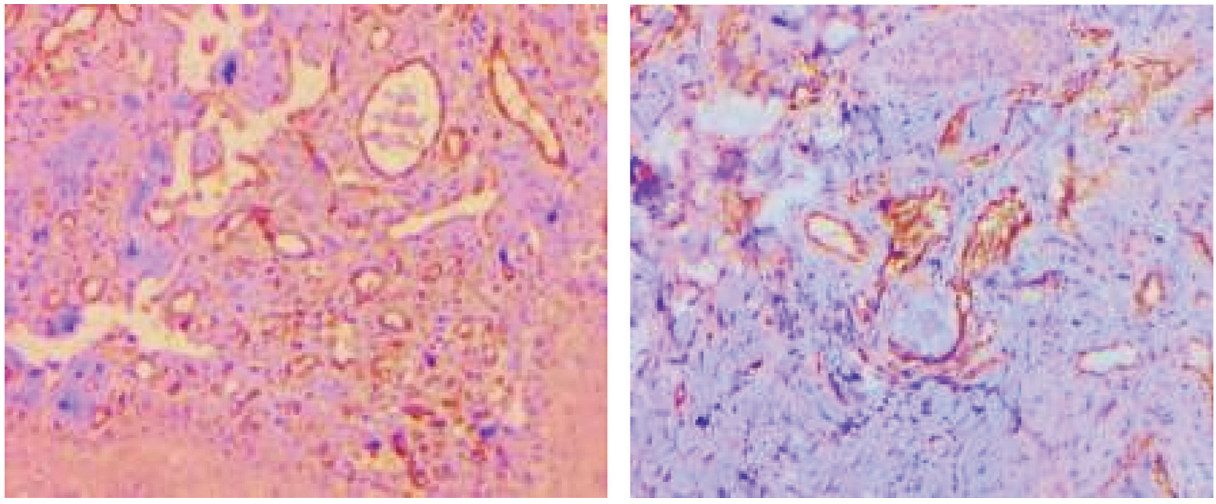

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Figure 3. Immunohistochemistry positive for (a) CD31 and (b) CD34.

single or multiple bluish red nodules or plaques that undergo secondary ulceration and bleeding [2]. Most cases occur spontaneously. Deeper soft tissue and visceral lesions can present with pain or discomfort. They are prone to develop locoregional recurrence and nodal and distant metastases, primarily hematogenously. Lungs are the most common site for metastases, and other sites include the liver, bones, soft-tissue structures, and lymph nodes [1,2].

Diagnosis is often delayed as the early lesions may resemble ecchymoses or hemangiomas [5]. The differential diagnoses include capillary haemangiomas, Kaposi's sarcoma, epithelioid hemangioendotheliomas, and hemangiopericytomas. The diagnosis can be made via skin biopsy and immunohistochemistry, positive for CD31, CD34, von Willibrand factor, and vascular endothelial factors [1].

Histologically, angiosarcomas are classified as well-differentiated or poorly differentiated. Numerous irregular vascular channels lined by endothelial cells are demonstrated in the well-differentiated variant, whereas spindle-shaped, polygonal, epithelioid, and primitive round cells with increased mitotic activity and poorly formed vascular spaces are seen in the poorly differentiated variant [6]. Other investigations such as ultrasonography, magnetic resonance imaging, computed tomography, and positron emission tomography imaging can be used to detect metastasis [1,2,6].

Localized angiosarcoma can be surgically excised with subsequent reconstruction. However, positive surgical margins are common after resection due to the extensive and rapid progression of the disease. Nonetheless, early and radical surgery with negative margins can offer the best prognosis. Most patients with head and neck angiosarcomas are not suitable candidates for surgical resection due to the close relationship of such tumors with underlying anatomical structures [1,2,6].

When surgical resection is not possible, immunotherapy or chemotherapy with agents like taxanes, doxorubicin, liposome doxorubicin, or ifosfamide is considered [3,4]. Metastatic angiosarcomas remain incurable and are often fatal [1,2]. The overall survival rate is about 50–60%, with a five-year survival rate of just 35% [1].

Conflict of interest: None declared.

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