

Erythematous plaque over the face

A 41-year-old male patient presented with gradually progressive painless erythematous swelling over the forehead and eyelids since nine months. The lesion appeared initially over the left temporal area. Examination revealed a dusky erythematous swelling on the forehead, upper and lower eyelids with a distinctly visible margin extending on the cheeks and behind the ear lobes

[Figure 1]. There was no tenderness, no mobility over underlying structures and no thickened cutaneous nerves. Sensations over the swelling were normal. Biopsy was taken from the lesion on the left temporal area. Histopathological features are shown in Figures 2, 3 and 4.

WHAT IS YOUR DIAGNOSIS?



Figure 1: Erythematous plaque on forehead and behind the ear

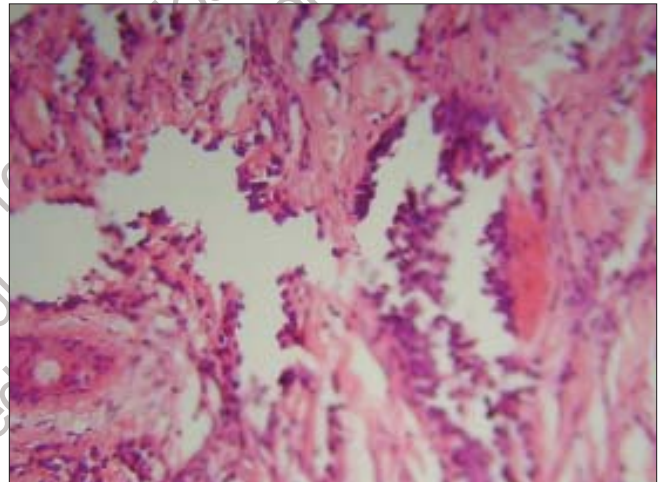


Figure 3: Skin biopsy high power (H and E, x200)

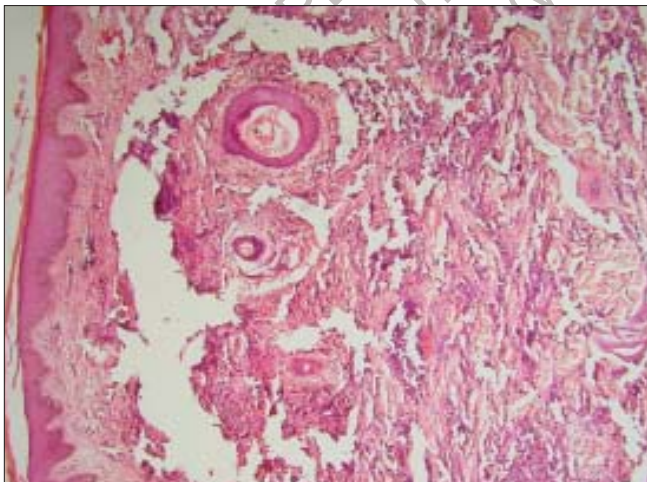


Figure 2: Skin biopsy low power (H and E, x40)

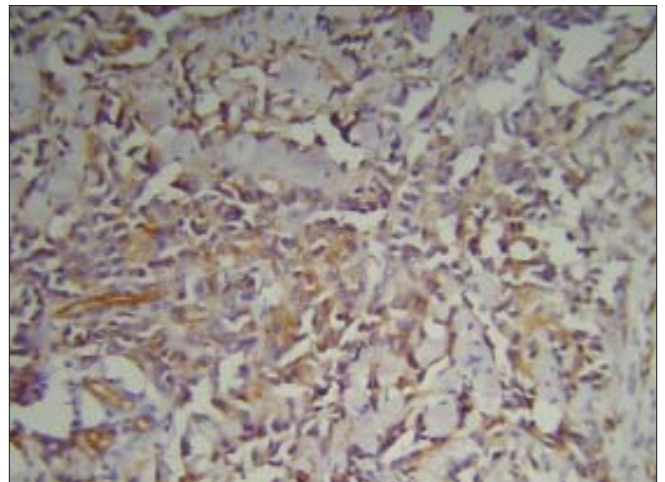


Figure 4: CD 34 staining of biopsy (x200)

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Diagnosis: Cutaneous angiosarcoma

Histopathology showed a non-epithelial neoplasm composed of irregular anastomosing vascular channels. The lining endothelial cells were large, polygonal, pleomorphic with hyperchromatic nuclei and moderate eosinophilic cytoplasm. Tumor was seen dissecting through the collagen. Mitotic activity was noted [Figures 2-3]. Immunohistochemistry revealed that CD 34 was strongly positive [Figure 4].

DISCUSSION

Angiosarcoma is a malignant tumor derived from the endothelium that occurs in a variety of anatomic sites including the skin. Usually, 60% arise within the skin or superficial soft tissues. Fifty per cent of cutaneous cases occur in the head and neck region and in particular, the scalp of elderly men.^[1] They are aggressive tumors that tend to recur locally and to metastasize despite aggressive multimodal therapy. Because of predilection of cutaneous angiosarcoma for multifocality and unapparent spread, complete surgical resection is often unattainable. Overall prognosis is poor with reported five-year survival of 10-35%.^[1]

The well-documented causative associations of angiosarcoma include chronic lymphedema of extremities, previous irradiation and persisting vascular malformation.^[1] Morgan *et al.* contended that cutaneous angiosarcoma is a disease of the elderly, aged between 60-80 years.^[1] In sharp contrast, our patient was 41 years old. The anatomic location of the lesion in our case was similar to the cases reported in their series. Ninety-six per cent of the cases of cutaneous angiosarcoma occur in the head and neck region. Excessive ultraviolet light exposure in this anatomic site has been invoked as a possible risk factor.^[1,2] Our patient had sun exposure time ranging between 6-8h per day for 20 years. Unusual vascular arrangements or density might also combine with UV light or thermal (heat) effect to potentiate oncogenesis.^[1]

Cutaneous angiosarcomas present in three different clinical settings: idiopathic cutaneous angiosarcoma of the head and neck, angiosarcoma complicating lymphedema and post-irradiation angiosarcoma.^[3] Low-grade angiosarcoma and early vascular lesion of Kaposi's sarcoma are difficult to distinguish from each other.^[3] Classical Kaposi's sarcoma has been reported to coexist with angiosarcoma.^[4] Occurrence of angiosarcoma in an area of telangiectasia in a case of systemic sclerosis has been reported.^[3] Angiosarcoma can mimic rhinophyma clinically.^[5] Cutaneous angiosarcoma can also present as an area of alopecia on the scalp, wherein focal scarring and follicular miniaturization was noted.^[6] Cutaneous angiosarcoma can metastasize to the lungs, liver, cervical lymph nodes, spleen and rarely, heart and brain.^[1] The mean time of survival after metastases was four months.^[1] Our patient has been referred for radiotherapy, as surgery was not considered feasible.

**S. P. Vijayalakshmi, Megha Arunharke*,
Sundaram***

Departments of Dermatology and *Pathology,
Nizam Institute of Medical Sciences, Hyderabad, India.

Address for Correspondence: Dr. S. P. Vijayalakshmi,
Department of Dermatology, NIMS, Hyderabad, India.
E-mail: kulvijaya@yahoo.co.in

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