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Angiosarcoma over Scalp: A Case Report and Review of Literature

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Abstract
Angiosarcomas are rare vascular malignancies, with a tendency to spread extensively both locally and systemically; present with metastasis in approximately 20% of patients. Bruise like patches on the face and scalp of elderly patients should raise the index of suspicion for angiosarcoma. We present a 56 year male patient with slowly widening induration, ulceration and oozing for 5 months on the scalp. The diagnosis of cutaneous angiosarcoma was made by histopathology of biopsy material taken from the involved area of the skin.

Introduction

Clinical presentation of angiosarcoma is variable, Ill defined bruise like areas or facial edema with minimal erythema are initial signs. Progressively more indurated plaques appear with nodular or ulcerated components. Other lesions that may need to be differentiated from angiosarcoma on the face and scalp include hemangiomas, Kaposi sarcoma, malignant melanoma, metastasis and vascular venous malformations.[1,2] Management includes a disciplinary team and may involve a combination of surgery, radiation and chemotherapy tailored to patient's age and associated co-morbidities.

Case Report

A 56 years old male presented with progressively extending, painless, ulcerated and red indurated skin lesions on left side of scalp in retro-auricular region for 5 months. Five months back, he noticed a bruise like lesion which he thought to be traumatic. Subsequently he noticed multiple dusky blue to reddish coloured nodular lesions in the same area. Some of the area ulcerated and black eschar formed over it. The mild ooze continued and the surrounding peripheral skin hardened.

On examination, the left side retro-auricular region had ill defined, indurated, non tender plaque of size 9×4cms with ulceration at places; there was decreased hair density over the lesion. The lesion was partially covered with black eschar

with areas of oozing and crusting "figure1". The anterior cervical and supraclavicular lymph nodes on left side were 2-3 cm in diameter, discrete, mobile, firm, and non-tender.

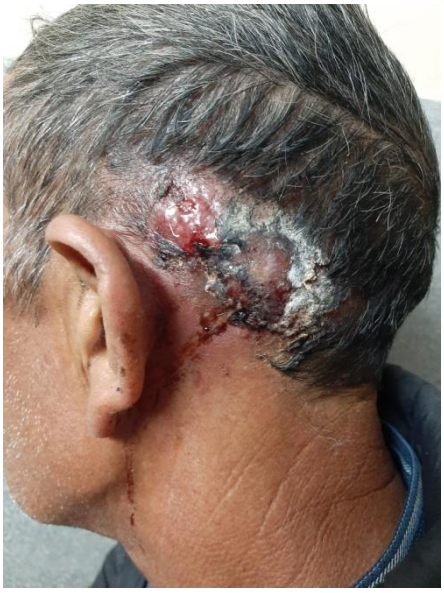

Figure 1: Lateral view of patient showing erythematous, indurated plaque with ulceration, partially covered with black eschar, with oozing and crusting.

Routine investigations including complete blood count, urine analysis, blood glucose, bleeding profile, liver and renal function tests; all were within normal limits. Skin biopsy from the edge of the plaque in the scalp divulged ulcerated epidermis with infiltration of entire reticular dermis by dense infiltrate of hypochromatic atypical cells that line irregular vascular channels and are seen to infiltrate and dissect the collagen. The histopathology was consistent with cutaneous angiosarcoma. X-ray and contrast-enhanced computed tomography (CECT) 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 for further management and advised regular follow-up.

Discussion

Angiosarcomas are rare malignant tumours derived from endothelial cells that line the blood vessels. Sporadic cutaneous angiosarcoma is commonly located on the scalp and upper part of face. Scalp angiosarcoma usually occurs in elderly men in the seventh to eighth decade of life and very rarely in sixth decade with an estimated male-to-female ration of 3:1.[3] Less than 5% cases of soft tissue sarcomas involve head and neck, 10% of which are angiosarcomas.[4] Predisposing factors for angiosarcoma include trauma, chronic lymphoedema, irradiation and age; however, most cases present with no obvious etiology.[5] Angiosarcomas has been classified into lymphoedema associated, radiation induced, primary breast angiosarcoma, sporadic cutaneous angiosarcoma (age related) and angiosarcoma of the soft tissue.[6] It was first described by Jones in 1964 as malignant angioendothelioma of skin.[7] Diagnosis is often delayed due to its variable presentations and benign appearance, which simulate a bruise or a hemangioma.

On histopathology, two main patterns of angiosarcomas are recognized: angiomatous and solid.[8] The angiomatous pattern is characterized by irregular anastomosing vascular channels that dissect through collagen, which was there in our patient. The blood vessels are lined by cells ranging from normal appearing endothelial cells to pleomorphic, hyperchromatic cells that exhibit multilayering. In solid form, tumor cells may be spindle or polygonal shaped with no or poorly identifiable vascular channels in poorly differentiated areas. The well differentiated angiomatous areas in angiosarcoma display cytological atypia, multilayering, papillary structures, and irregular vascular channels.[9]

Generally radical surgery and post operative radiotherapy are advocated to treat these tumours. Extensive microscopic spread of angiosarcoma result in difficulty achieving negative margins after surgery; so microscopic control of surgical margins may have a role in guiding extend of tumour resection.[10,11] Chemotherapeutic agents

such as doxorubicin, cyclophosphamide, methotrexate, dacarbazine, vincristine and actinomycin D are used in case systemic angiosarcoma. Systemic paclitaxel therapy showed encouraging results.[12,13]

Angiosarcoma tend to have aggressive local invasion and distant metastasis through lymphatics and hematogenous route. Regional lymph node involvement occurs in 20-30% cases, which is higher than most other sarcomas.[2,5] The most common site for distant metastasis is lungs followed by liver, spleen and bone. Prognosis of angiosarcoma is poor because of its high potential for metastasis. The five year survival is 10%-20%.[4,9] Tumour of more than 10cm, greater depth of invasion, delay in diagnosis due to initial misinterpretation and high mitotic count are bad prognostic factors reported in cutaneous angiosarcomas.[9,14]

Conflicts of interest: Nil

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