An Uncommon Profile of a Case of Angiosarcoma of Scalp and Face

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Introduction

Angiosarcoma is a rare soft tissue sarcoma usually seen in the head, face and neck [1]. Head and neck angiosarcomas are seen in elderly population with the mean age greater than 60 years [2]. Angiosarcoma also has a slight predilection for Caucasian patients and the male gender. The estimated ratio of male-to-female predominance is approximately 3:1. Angiosarcoma involving the scalp of old patients was first described as a distinct subgroup by Wilson-Jones and is usually limited to the skin and soft tissues. The disease occurs in the dermis and presents as single or multiple bluish or red nodules or plaques which ulcerate or bleed. Metastasis to regional lymph nodes or lungs can occur [4].

Case Report

A 59 year old lady presented with complaints of raised dark coloured gradually progressive lesion on scalp and left side of face of 2 years duration associated with a sensation of intense itching and crawling of insects. Pruritus was disabling with disturbance of sleep. There was history of multiple ulcerations over scalp associated

with profuse blood tinged discharge and hair loss since 6 months. There was no history of chronic cough, breathlessness, weight loss, fever, oral or genital ulcers, rashes, trauma, insect bite or preexisting skin disorders. She was previously evaluated at various hospitals with multiple skin biopsies but no definite diagnosis was arrived at. A trial of antipsychotic medications as a case of delusional parasitosis provided no relief.

Dermatological examination revealed a large lesion of size 30×15 cm involving whole scalp leaving a small area in frontal region and extending on to the left side of face with lobulated polycyclic margins. Few superficial ulcers of varying sizes ranging from 3×2 cm to 8×6 cm were seen over the occipital and post-auricular region with profuse serosanguinous discharge (Fig. 1). Multiple ill-defined areas of scarring alopecia were seen with shiny atrophied skin in between the ulcerated areas. The patient subsequently developed multiple ulcerations over scalp with increase in discharge. A provisional diagnosis of lupus vulgaris of scalp was entertained on clinical grounds, pending histopathology and standard anti tubercular therapy exhibited though Mantoux test was negative. However the ulcerations persisted. Other differential diagnoses considered were follicular mucinosis, mycosis fungoides and adnexal tumours.

Routine hematological and biochemical investigations were all normal. Pus swab for bacterial and fungal cultures were negative. MRI revealed skin, subcutaneous tissue and adjoining subcutaneous fat thickening on left side of face and scalp. An incisional wedge biopsy from the lesion revealed cells with round to oval vesicular nuclei with prominent nucleoli arranged singly and in small nests and

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Fig. 1 Lesion involving the scalp with multiple ulcerations and extending onto the face

at places around vascular lumina. Intracytoplasmic vacuolations and scattered mitotic figures were seen. The tumour cells were seen infiltrating reticular and papillary dermis (Fig. 2).

IHC for CD31 marker using Dako reagent and JC 70A clone was positive (++) in tumour cells (Fig. 3). IHC for other markers like CA, CD3, CD20, S100, HMB45, CK, CD68, SMA, CD8 were negative. Based on the clinical features, histopathological picture and immunohistochemistry positivity for CD31, a diagnosis of Dermal Malignant Haemangioendothelioma (low grade angiosarcoma) was made.

The patient was further subjected to a whole body PET scan for staging and detection of metastasis. The PET scan revealed diffuse nodular thickening of left frontal, left temporal, bilateral parietal and occipital scalp. No calvarial erosion was seen. A few enlarged bilateral parotid, level V and right level II cervical lymphadenopathy was noted. There was no evidence of any distant metastasis. FNAC of the lymph node revealed acute inflammation.

Fig. 2 Biopsy of the lesion from scalp H&E stained 40×. Vascular spaces lined by endothelial cells showing nuclear atypia and intracytoplasmic inclusions

As the lesion was locally advanced and not amenable for R0 resection, a joint decision was made to excise the involved scalp and facial lesions as far as resectable and then give her adjuvant therapy.

Fig 3 IHC positive for CD31 (10×)

She underwent surgery by a joint team of surgical oncologists and reconstructive surgeons. The pericranium was not infiltrated and hence not excised. A split thickness skin graft taken from thigh was used to cover the excised area (Fig. 4). Post op recovery was excellent with 100 % graft take (Fig. 5). Histopathology of the excised specimen revealed all margins to be positive. She is being administered adjuvant radiotherapy and chemotherapy and will be placed on follow up.

Discussion

Sarcomas are uncommon in the head and neck region constituting only 1 % of all malignant neoplasms in this region [5]. Angiosarcomas make up less than 1 % of all sarcomas. Angiosarcoma of the scalp occurs in elderly male patients 68 to 76 years old with an overall male-to-female ratio of 3:1 [1, 3]. Our patient however was a female in the late fifties.

Cutaneous angiosarcoma of the head and neck is a distinct subtype which most commonly presents as an enlarging purple bruise-like lesion that develops over several months [6]. Ulcerated, fungating and haemorrhagic lesions indicate advanced disease. It is often multifocal and associated with a high incidence (10–15 %) of lymph node metastasis [7]. Distant metastasis may occur in up to 50 % with

Fig 4 Excision and grafting

Fig. 5 Graft take-up 2 weeks post op

the lung being the most common site followed by liver [8]. Younger patients have better prognosis while presence of metastasis at the initial presentation is associated with poor prognosis. Overall prognosis is reported to be very poor, the five-year survival being less than 10–30 % [3].

Surgery is considered the mainstay of treatment with wide local excision and frozen section control. The treatment described is wide excision of the lesion to achieve histologically tumour-free margin as this has a direct impact on the prognosis [3, 8]. Prognosis correlates well with the ability to attain clear surgical resection margins although the tendency of scalp angiosarcoma to demonstrate a diffuse pattern of clinically undetectable spread makes resection challenging [9]. The reported outcomes of radiotherapy alone have largely been unsatisfactory. Chemotherapy has been suggested for unresectable cases, but has generally not proven beneficial [10]. Local recurrence and metastases are very frequent regardless of the treatment modality employed.

Aim of reporting this case is its rarity and to highlight the multidisciplinary involvement in the diagnosis and management of such a case. IHC is a helpful tool to confirm diagnostic suspicion. Another unique symptom in this case, not reported in literature, was intense pruritus with sensation of crawling of insects not relieved by antihistamines.

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