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Case Report

Rare case of recurrent angiosarcoma of the scalp: a case report and review of literature

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ABSTRACT

Angiosarcoma is a rare soft tissue tumour and forms less than 1% of all sarcomas. It usually occurs in the head and neck, and lesions in the scalp are seen in elderly people. Clinically, it varies from a small plaque to multifocal nodules and ulcers. The treatment varies with the extent of the disease. Most cases are treated with wide excision with reconstruction. Radiotherapy and chemotherapy is advised in the recurrent or extensive lesions with regional or distant metastasis. Recently, immunomodulation have been tried. Authors present a rare case of a 43-year-old female patient having bleeding scalp nodular lesions with ulcers. There was past history of nodular lesion over same area on scalp 17 years back which was diagnosed as angiosarcoma and was treated. On biopsy ulcerative nodular lesions were diagnosed as angiosarcoma. Wide local excision with craniectomy was done. Reconstruction using a latissimus dorsi flap was done at a second stage. The patient was not given postoperative radiotherapy or chemotherapy. There has been no recurrence for two years of follow-up.

Keywords: Angiosarcoma, Vascular neoplasms, Hemangiosarcoma, Sarcoma

INTRODUCTION

Angiosarcoma is a rare vascular tissue sarcoma usually seen in the head, face and neck. Angiosarcoma comprised of around 10% of the soft tissue sarcomas involving head, face and neck.[4,5,9] Angiosarcoma of the scalp is relatively more common in Caucasian elderly males. Angiosarcomas present as single or multiple bluish or red nodules or plaques which ulcerate and bleed. Metastasis to regional lymph nodes or lungs is common. Around 10% of these lesions develop in patients with chronic lymphedema also called as Stewart Treves syndrome. It is also noted in patients exposed to radiation.[9,13] Microscopically, the tumour can vary from differentiated to poorly differentiated types and may occasionally involve deeper structures. Treatment of these lesions is usually by radical excision and subsequent reconstruction.[3,9] Radiotherapy and chemotherapy is used tumours with involved margins, in unresectable tumours and those with distant metastasis. Angiosarcoma has high recurrence rate. Prognosis is poor with 5-year survival being 10-30%.

CASE REPORT

A 43-year-old female patient presented to the outpatient department with multiple nodules with ulcers over right occipito-parietal region of scalp which bleeds on touch. It started as multiple small nodules which increased in size over last 3 months and ulcerated in last 15 days. There was past history of being operated 17 years back for similar nodular swelling in the scalp and was diagnosed as angiosarcoma. Wide local excision with local flap

repair was done at that time. Patient lost for follow-up at that time.

On examination, there was 8 cm × 7 cm × 7 cm ulcerative nodular lesion over the right occipito-parietal region of the scalp (Figure 1). It was brownish red in colour, firm in feel with irregular indurated margins and bleeds on touch. The rest of the scalp was normal. There were no palpable lymph nodes in the neck. Biopsy was performed and the specimen sent for examination. Histology reported dissecting and anastomosing vascular channels with focal cytological atypia suggestive of malignancy in the dermis present beneath ulcerated and inflamed squamous epithelium. Necrosis was also noted. Histologically, features were consistent with a neoplasm of vascular tissue involving the dermis, a cutaneous angiosarcoma of the scalp. Metastatic work-up of the patient was done. No distant metastasis was noted. CT scan head showed erosion of outer table of skull in parietal region underlying the ulcer.

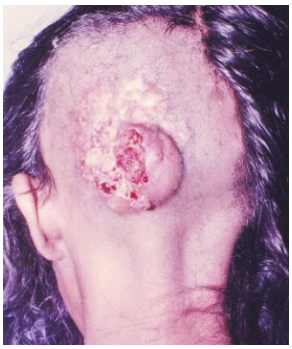

Figure 1: Clinical photograph of recurrent angiosarcoma of scalp.

Wide local excision with local craniectomy was performed (Figure 2 and Figure 3). Histopathology of specimen showed free margins. Then, the defect was closed with Latissimus Dorsi (LD) flap with microvascular reconstruction in second stage operation (Figure 4). Patient was started on doxorubicin-based chemotherapy but it was withdrawn after first cycle as patient developed severe neutropenia. No radiotherapy was given to the patient. Patient is on regular follow-up since two years and there is no local recurrence or distant metastasis.

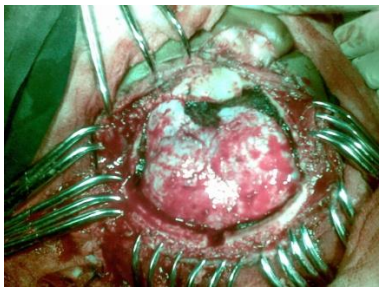

Figure 2: Intraoperative photograph of angiosarcoma scalp.

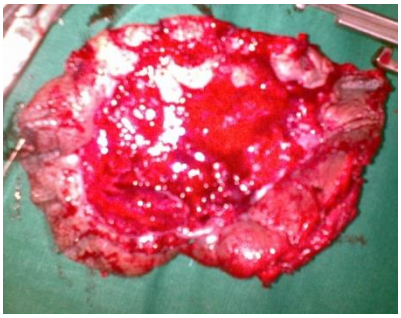

Figure 3: Excised specimen.

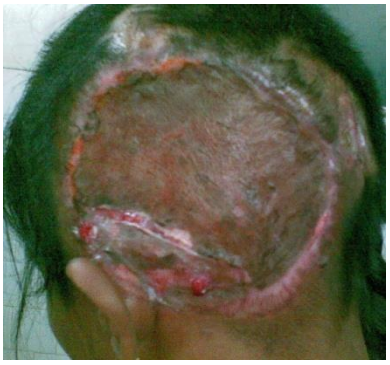

Figure 4: Clinical photograph: post-operative period after reconstruction.

DISCUSSION

Angiosarcoma is a rare vascular tissue tumour arising from the endothelial cells with histology varying from well-differentiated tumours to poorly differentiated ones and form less than 1% of all sarcomas.[3] Low-grade angiosarcomas are well differentiated while the high-grade lesions are poorly differentiated and consist of sheets of pleomorphic cells with areas of haemorrhage, disordered architecture, cells with hyperchromatic and pleomorphic nuclei with prominent mitotic activity. Both types are associated with extensive local growth. The high tendency to metastasis is associated with the absence of vascular endothelial cadherin (VE - cadherin), as the vascular endothelial cadherin is seen in the normal endothelium margins.[13] No correlation was found between grade of angiosarcoma and survival.[3,9] The cutaneous angiosarcoma following treatment of breast carcinoma was attributed to chronic lymphedema also known as Stewart-Treves syndrome. In breast-conserving surgery the incidence of lymphedema is reduced. A form of cutaneous, post-radiation angiosarcoma of the breast (CPRSAB) is also described.[1] The basis of development of angiosarcoma in these patients has been postulated to be radiation-induced connective tissue damage.[2] Exposure to the sun with actinic skin damage was proposed as a cause of angiosarcoma due to its predominance in Caucasians and rarity in the coloured races but this theory was refuted by the fact that most patients with angiosarcoma of the scalp had hair which gives protection from exposure to the sun.[7] Other

predisposing factors that are reported include occurrence of tumour in previous arterio-venous fistula, herpes zoster sites, telangiectatic nevus, chronic osteomyelitis, other vascular-lymphatic abnormalities and exposure to arsenic, thorotrast and polyvinyl chloride.[3,5,9] In most patients like ours no underlying predisposing factor is found. Angiosarcoma of the scalp occurs in elderly male patients 70 to 80 years old with an overall male-to-female ratio of 2:1.[5,9]

Most patients present with a bruise-like macules or a non-bruise-like nodules.[9] However, its presentation can vary as indurated, erythematic nodules, fungating masses, ulcerations or sometimes as bleeding lesions, as in our patient. Ulcerated, fungating and haemorrhagic lesions indicate advanced disease. A diffuse widespread angiosarcoma of the scalp presenting with Kasabach-Merritt phenomenon with consumption coagulopathy and thrombocytopenia is reported which resolved only with the regression of the tumour.[7] Also a rare case where the angiosarcoma of the scalp presented as extensive scarring alopecia is also reported in literature.[8] Angiosarcomas has the highest rate of lymph node metastases among all soft tissue sarcomas of the head and neck and distant metastasis may occur in up to 50% with the lung being the most common site followed by liver.[6] Delayed recurrence of angiosarcoma at the distant sites has been reported for which regular, lifelong surveillance is required. In our case recurrence occurred after 17 years at same site. In soft tissue sarcomas, the size of the tumour, its grade and depth including neurovascular or bone involvement are the predictors of metastasis. Therefore early diagnosis and aggressive management of angiosarcoma is needed [6]. Recurrence is more common and early in multifocal disease.[9] Overall prognosis is reported to be very poor, the five-year survival being less than 10-30%.[9]

Wide local excision of the lesion to achieve histologically tumour-free margin is treatment of choice in these patients microscopic spread of the tumour is extensive.[4,6,9] Primary closure of the wound is often not possible after wide excision and a staged reconstruction is performed only after confirming tumour-free margins on histopathological examination. The reconstructive options available are split-thickness skin grafts, local flaps and free flaps. The skin grafts are commonly used except when the excision includes the pericranium or when there is history of previous radiotherapy. Local rotation flap is indicated when the pericranium is also excised and the defect is not extensive.[14] When the angiosarcoma is multicentric and extensive, wide excision of almost whole scalp is required.[14] Such an extensive defect is reconstructed with free flaps. Postoperative low-dose radiation is effective in treating local recurrence and in diffuse multifocal lesions.[7] Improved survival rate with routine use of postoperative radiotherapy is seen in only 21% patients who had tumour-free margins.[9] Use of high-dose brachytherapy with a surface mould technique in extensive angiosarcoma of the scalp has been reported.[8] It reduces the risk of recurrence at margins. Recently immunomodulation with cytokine therapy have been reported in the form of intralesional interferon alpha-2b and interleukin-2 combined with surface radiotherapy, as an alternative to surgery.[11] No established chemotherapy regimens are available for angiosarcoma of scalp. The use of liposomal doxorubicin with radiotherapy has been reported.[12] Recently paclitaxel is also tried in the treatment of angiosarcomas.[13]

CONCLUSION

Angiosarcoma of the scalp is an extremely aggressive vascular tumour with poor prognosis which can be improved with early diagnosis and aggressive early treatment. In all elderly patients with lesions on the scalp, head and neck biopsy should be performed for early detection of tumour. Complete metastatic work-up is must while planning the treatment of angiosarcoma scalp. Wide local excision to achieve tumour-free margin is associated with improved survival. Widespread lesions with margin involvement other treatment modalities as described is advised. Lifelong follow-up is advised to detect any delayed distant metastasis and local recurrence.

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