ANGIOSARCOMA OF SCALP: A CASE REPORT

Pallavi Goyal[1], Savita Agrawal[2], Manisha Nijhawan[3]

[1]Junior Resident, Department of Skin & VD, Mahatma Gandhi Medical College & Hospital, Jaipur.
[2]Assistant Professor, Department of Skin & VD, Mahatma Gandhi Medical College & Hospital, Jaipur.
[3]Professor & HOD, Department of Skin & VD, Mahatma Gandhi Medical College & Hospital, Jaipur.

ABSTRACT

Cutaneous angiosarcoma is relatively rare vascular tumour which constitute less than 1% of all malignant mesenchymal tumours. It usually occurs in the head and neck, especially in the scalp, in elderly people. Its presentation varies from a small plaque to multifocal nodules.[1,2,3,4] We present a case of an elderly male presenting with progressive non-healing ulcer over scalp of 9 months duration. Histological examination of the biopsy revealed features of an angiosarcoma, with positive markers, vimentin and CD 31.

KEYWORDS

Angiosarcoma, Vascular Tumour, Soft Tissue Sarcoma.

HOW TO CITE THIS ARTICLE: Goyal P, Agrawal S, Nijhawan M. Angiosarcoma of scalp: a case report. Journal of Evolution of Research in Dermatology and Venereology 2016; Vol. 2, Issue 1, Jan-June 2016; Page: 6-7.

INTRODUCTION

An angiosarcoma (AS) is an uncommon malignant neoplasms characterized by rapidly proliferating, extensively infiltrating anaplastic cells derived from blood vessels and lining irregular blood-filled spaces. Approximately 50% of angiosarcomas occur in the head and neck, but they account for less than 0.1% of head and neck malignancies.[5] Cutaneous angiosarcoma is more frequent in males than in females, with a male-to-female ratio of 2:1.[6] Cutaneous angiosarcoma of the head and neck tends to occur in the elderly population.[1,2] 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.[7,8]

CASE REPORT

A 53-year-old male presented with large non-healing ulcer over the scalp for past 9 months. It started as a small nodular lesion which progressed rapidly and increased in size to involve the parietal & occipital region of the scalp. In due course multiple bleeding ulcers developed within the lesion. Lesion was painless to start with but now became painful for past 3 months. His general physical examination and systemic examinations were normal. Cutaneous examination revealed large, multifocal, erythematous plaque of size 15x12 cm with multiple ulcers and crusting. Lesion was non-tender, firm and bleeds on touch.

Fig. 1: Erythematous plaque with multiple bleeding ulcer

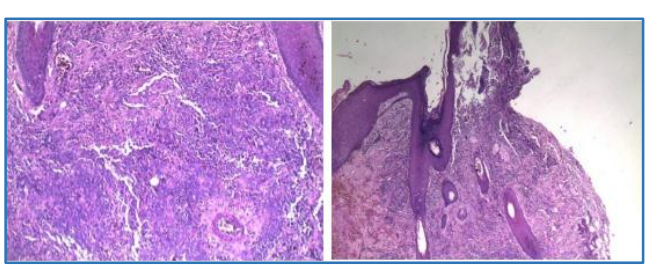

Fig. 2

Investigations revealed normal blood counts, blood glucose levels, liver and renal function test. Biopsy from lesions shows a non-epithelial neoplasm involving whole of the dermis in a diffuse fashion. [Figure 2] The neoplasm is made up of numerous variously sized and shaped vascular channels some of which are dilated in the upper dermis. The neoplastic cells line these vascular channels and show variable morphology some being elongated and spindle shape while others are more plump, polygonal and some even plasmacytoid. Cords and nodules of solid proliferation of these cells are also seen. The neoplastic cells show moderate to marked nuclear pleomorphism with scattered normal and abnormal mitotic figures. Overlying epidermis is hyperplastic.

Financial or Other, Competing Interest: None.
Submission 22-04-2016, Peer Review 21-05-2016,
Acceptance 27-05-2016, Published 15-06-2016.
Corresponding Author:
Dr. Pallavi Goyal,
PG Hostel 4 A,
Mahatma Gandhi Medical College & Hospital,
Sitapura, Jaipur.
E-mail: drpallavigoel@gmail.com

The neoplasm reaches the base and both lateral margins Suggestive of angiosarcoma.

The presence of distal metastasis was ruled out by an chest X-ray and abdominal ultrasound.

Marker study was positive for vimentin & CD31, suggestive for angiosarcoma.

DISCUSSION

Angiosarcoma is a rare soft tissue sarcoma usually seen in the head, face and neck. 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. Cutaneous angiosarcomas may be associated with chronic lymphoedema, previous radiation therapy, treatment of breast carcinomas and immunosuppression in renal transplant patients.[9,10,11]

Most patients present with a bruise-like macule or a non-bruise-like nodule.[7] Other common presentations include indurated, erythematic nodules, fungating masses, ulcerations or sometimes as bleeding lesions, as in our patient. Ulcerated, fungating and haemorrhagic lesions indicate advanced disease while unusual presentations too have been reported.

It 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. [12]

In one of the largest series to date, Perez et al reported on 88 patients with cutaneous angiosarcoma divided into 4 groups: 38 head and neck, 30 radiotherapy-induced, 13 sporadic trunk/extremity, and 7 Stewart-Treves syndrome related. The median age at diagnosis for the entire cohort was 70 years (72.5 y for head and neck), with 95% being white. Of those with head and neck angiosarcoma, 82% were male. Perez et al observed only 7% of patients to have regional nodal involvement.[13]

Younger patients have better prognosis,[7,14] 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%.

REFERENCES

1. Young RJ, Brown NJ, Reed MW, et al. Angiosarcoma. Lancet Oncol 2010;11(10):983-91.
2. Mark RJ, Poen JC, Tran LM, et al. Angiosarcoma a report of 67 patients and a review of the literature. Cancer 1996;77(11):2400-6.
3. Ogawa K, Takahashi K, Asato Y, et al. Treatment and prognostic of sarcoma of the scalp and face: a retrospective analysis of 48 patients. Br J Radiol 2012;85(1019):e1127-33.
4. Maddox JC, Evans HL. Angiosarcoma of the skin and soft tissue: a study of forty four cases. Cancer 1981;48(8):1907-21.
5. Sturgis EM, Potter BO. Sarcomas of the head and neck region. Curr Opin Oncol 2003;15(3):239-52.
6. Meis-Kindblom JM, Kindblom LG. Angiosarcoma of soft tissue: a study of 80 cases. Am J Surg Pathol 1998;22(6):683-97.
7. Paulik TM, Paulino AF, Mcginn CJ, et al. Cutaneous angiosarcoma of the scalp: a multidisciplinary approach. Cancer 2003;98(8):1716-26.
8. Wollina U, Fuller J, Graefe T, et al. Angiosarcoma of the scalp: treatment with liposomal doxorubicin and radiotherapy. J Cancer Res Clin Oncol 2001;127(6):396-9.
9. Aust MR, Oslen KD, Lewis JE, et al. Angoisarcoma of the head and neck, clinical and pathological characteristics. Ann Otol Rhinol Laryngol 1997;106(11):943-51.
10. Billings SD, McKenney JK, Folpe AL, et al. Cutaneous angiosarcoma following breast conserving surgery and radiation: an analysis of 27 cases. Ann J Surg Pathol 2004;28(6):781-8.
11. Brenn T, Fletcheer CD. Radiation associated atypical vascular lesions and angiosarcoma: clinicopathological analysis of 42 cases. Am J Surg Pathol 2005;29(8):983-96.
12. Obeng MK, Henandez A, Dastgir A, et al. Angiosarcoma of the scalp with calvarium involvement in a 50 year old African-American man. J Nat Med Assoc 2004;96(11):1507-12.
13. Perez MC, Padhya TA, Messina JL, et al. Cutaneous angiosarcoma: a single-institution experience. Ann Surg Oncol 2013;20(11):3391-7.
14. Sondak VK, Recs R, Paulik T, et al. Emedicine skin: angiosarcoma: surgical and nonsurgical treatment. Medscape-http://emedicine.medscape.com/article/1296442-overview.