Cutaneous Angiosarcoma - A Clinicopathological Study of 16 Cases over a Period of Nine Years

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ABSTRACT

BACKGROUND

Cutaneous angiosarcoma is an aggressive malignant mesenchymal vasoformative neoplasm accounting for 1 % of all soft tissue sarcomas. It carries a poor prognosis with high local recurrence rate. Cutaneous angiosarcoma occurs in 3 clinical settings namely idiopathic angiosarcoma of head and neck, angiosarcoma associated with chronic lymphedema and post irradiation angiosarcoma. Idiopathic angiosarcoma of head and neck of elderly are distinct from other types of angiosarcoma. Clinicopathological correlation of cutaneous angiosarcomas diagnosed during a period of nine years from January 2010 to July 2019 was done.

METHODS

We retrospectively studied and analysed specific data such as age, sex, site, size, clinical setting as well as six histopathological parameters and follow up.

RESULTS

There were 30 patients diagnosed as angiosarcoma. 16 of these cases were cutaneous. Their mean age was 60 years (44 – 80 years). Mean size of cutaneous tumours was 4 cms (3 – 10 cm). 15 were primary; one had history of preceding lymphedema. Most common location was scalp (14) followed by leg (2). Predominant pattern was vasoformative (75 %) which included spindle (62.5 %) and epithelioid (37.5 %) cell types. Four (25 %) showed extensive necrosis; ten (66.6 %) had high mitotic index and 12 (75 %) showed tumour infiltrating lymphocytes. All cases (100 %) were positive for CD31 & CD34. All underwent wide local excision. Adjuvant chemotherapy was given in two cases and adjuvant radiotherapy in three cases. Of the seven cases with follow up, five cases died of their disease and two survived.

CONCLUSIONS

Larger tumour size & older age group were associated with worse outcome. All our 14 cases very well fit with the angiosarcoma of scalp & face in elderly persons as described in the literature. This disease has unique characteristics compared to angiosarcoma in other locations.

KEYWORDS

Cutaneous, Angiosarcoma, Scalp, Elderly

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BACKGROUND

Cutaneous Angio-Sarcoma (AS) is an aggressive malignant mesenchymal vasoformative neoplasm accounting for 5 % of malignant skin tumours and less than 1 % of all sarcomas.[1] They carry a poor prognosis with high local recurrence rate. Angiosarcomas can occur at any site in the body, most common site being head and neck followed by breast, viscera and bones. Cutaneous angiosarcoma occurs in 3 clinical settings namely idiopathic angiosarcoma of head and neck, angiosarcoma associated with chronic lymphedema and post irradiation angiosarcoma. Idiopathic angiosarcoma of head and neck of elderly are distinct from other types of angiosarcoma. The clinical course includes local recurrences followed in some cases by pulmonary and lymph node metastasis. Differential diagnosis for cutaneous angiosarcoma includes haemangioma in better differentiated lesions, Kaposi sarcoma for those with predominantly spindle cell component and carcinoma or amelanotic melanoma for poorly differentiated types. It's important for the pathologist to give a correct diagnosis, as misdiagnosis may result in inappropriate management and incorrect assessment of prognosis.

This study has been conducted to understand the clinicopathological features, treatment response and clinical outcomes of cutaneous angiosarcoma.

METHODS

This is a retrospective study, analysing all patients who were histologically proven as angiosarcoma in our department between the period of January 2010 to July 2019.

After obtaining the necessary approvals from the Institute Research Committee and the Institute Ethics Committee, we recorded clinical and histopathological parameters for each of the 16 patients including demographic features (age, sex, location), anatomic location, size, type (primary, postradiation, lymphedema associated), histologic pattern [vasoformative / solid / mixed], predominant cell type [epithelioid / spindle], presence / absence of necrosis, mitosis, tumour infiltrating lymphocytes, immunohistochemical stains [CD31 & CD34] and follow up.

All of the necessary information and relevant images were obtained from the institute medical records library and photo archives of the Department of Pathology, Dermatology & Radiotherapy.

RESULTS

A total of 30 cases of angiosarcomas were diagnosed in our department during this nine-year study period. Among those, 16 cases were cutaneous angiosarcoma with an equal distribution of males and females aged between 44 – 80 years (mean = 60.4 years) with tumour size ranging from 3 – 10 cm (mean = 4 cm). Vast majority (14 cases) of the cases were primary angiosarcoma of head and neck of elderly and the remaining two cases were angiosarcoma of lower limb (Table I, Figure 1, 2) of which one was preceded by chronic filarial lymphedema. All 16 patients underwent wide local excision (Figure 3) with adjuvant chemotherapy (Ifosfamide and Adriamycin) for two cases. Of the seven patients with follow up, five died due to their disease despite receiving multiple cycles of chemo radiation and two cases survived. Among the cases who survived, one was patient with angiosarcoma following chronic filarial lymphedema. Histologically (Table II), 12 cases (75 %) were of high grade and four of low grade. Predominant pattern was vasoformative (66.67 %), four had solid pattern and two remainder cases showed mixed pattern. Cell morphology showed both spindle (62.5 %) and epithelioid (37.5 %) cell types. Four (25 %) showed extensive necrosis, ten (66.6 %) had high mitotic index and 12 (75 %) showed tumour infiltrating lymphocytes (Figure 4, 5). All cases (100 %) were positive for CD31 & CD34 (Figure 6).

Significant clinical and histopathological patterns have been summarized in Table I, II.

	Clinical Types	Nos.
1	AS of head & neck of elderly	14
2	AS associated with chronic lymphedema	2
3	Post irradiation AS	0
	Total	**16**

Table 1. Clinical Types

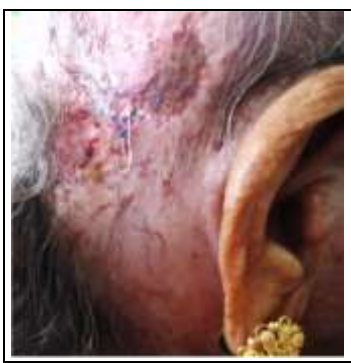

Figure 1. 69 / F Presented with Multiple Violaceous Papules

Figure 2. 61 / M with Angiosarcoma Secondary to Chronic Filarial Lymphedema

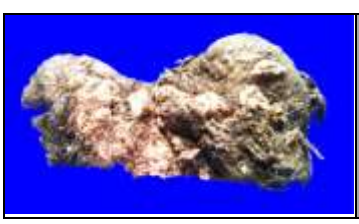

Figure 3. Wide Local Excision Specimen Shows Multiple Induratednodules with Ulceration Measuring 10 x 8 x 5 cms

Figure 4. Cut Section is Haemorrhagic

Grade	High Grade	75 %
	Low Grade	25 %
Pattern	Vasoformative	66.67 %
	Solid	25 %
Cell type	Spindle	62.5 %
	Epithelioid	37.5 %
IHC	CD31	100 %
	CD34	100 %

Table 2. Histologic Features

	Our Study 11 Cases (9 yrs.)	Morgan et al 47 Cases[5]	Pawlik et al 29 Cases (27 yrs.)[6]
Age	60.4 yrs.	75.1 yrs.	71 yrs.
M:F	1:1	36:11	18:11
Size	4 cm	5.3 cm	4 cm
Multiple	7 / 16	-	12 / 29
High Grade	75 %	-	76 %
Low Grade	25 %	-	24 %
Vasoformative	66.67 %	64 %	70 %
Solid	25 %	25 %	30 %
Mixed	8.33 %	21 %	0

Table 3. Comparison with Studies in Literature

DISCUSSION

Cutaneous AS is an extremely rare tumour as obvious from the result that only 16 cases were found over nine yrs. Overall, it has a strong predilection to elderly males who present with erythematous oedema on head and neck area called idiopathic primary angiosarcoma of head and neck of elderly (Figure 1), also known as Wilson-Jones angiosarcoma. In our study the mean age was 60 years with an equal sex distribution. The most common site in our study was scalp, as would be expected. Idiopathic cutaneous angiosarcomas of head and neck of elderly are distinct from angiosarcomas in other locations.[2] Their increased risk for multiplicity, recurrences, metastasis to brain parenchyma and propensity to spread microscopically far beyond the grossly visible margins make the achievement of surgically free margins a daunting task.[3] Older age group and larger size are particularly associated with grave prognosis.[4]

Post radiation AS is on the rise because breast cancers are being treated with radiation as opposed to radical mastectomy. But none of our cases could be attributed to prior radiation. Lymphedema associated AS, also called Stewart-Treves syndrome, is the least common form of the disease. Chronic lymphedema is one of the identified risk factors in angiosarcoma, with proteins in lymph fluid inducing immunosuppression & promotion of vascular oncogenesis. One of our cases that involved the extremity was a 61-year-old male with filarial lymphedema associated angiosarcoma (Figure 1). A wide variety of histopathological features have been described, the most characteristic being anastomosing vessels lined by atypical neoplastic spindle or epithelioid cells dissecting through the collagen bundles. High grade lesions show an undifferentiated solid pattern, increased mitosis and extensive necrosis as described in FNLCC grading of soft tissue sarcomas. Table III shows comparison with studies in literature

Limitations

The limited number of cases in our study restricted us from doing statistical workup.

CONCLUSIONS

Despite the limited number of cases in our study, which restricted us from conducting statistical work up, it was found out that tumour size (larger) & age (older) were associated with worse outcome. All our 14 cases very well fit with the angiosarcoma of scalp & face in elderly persons as described in the literature. This disease has unique characteristics compared to angiosarcoma in other location.

Data sharing statement provided by the authors is available with the full text of this article at jebmh.com.
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