Cutaneous angiosarcoma – a rare malignant vascular tumour

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Abstract

Angiosarcoma is a rare malignant vascular tumour arising from the both vascular and lymphatic epithelium. It accounts for less than 1% of all sarcomas. It usually arises in the scalp or face and is usually locally advanced at presentation[1]. It is more common in elderly men with an age peak of 60-70 years[2].

It occurs in 3 settings as idiopathic angiosarcoma of the face, scalp, and neck, angiosarcoma associated with chronic lymphoedema (Stewart Treves syndrome) or post-irradiation angiosarcoma[3].

Cutaneous angiosarcoma has a poor prognosis with a 5-year survival rate ranging from 26-51%[4]. Large tumour size, high tumour grade, large number of mitosis and older age are associated with poor prognosis[5].

Here we report 70-year-old previously healthy lady who presented with erythematous tender nodules in front of the L/Ear for one year duration.

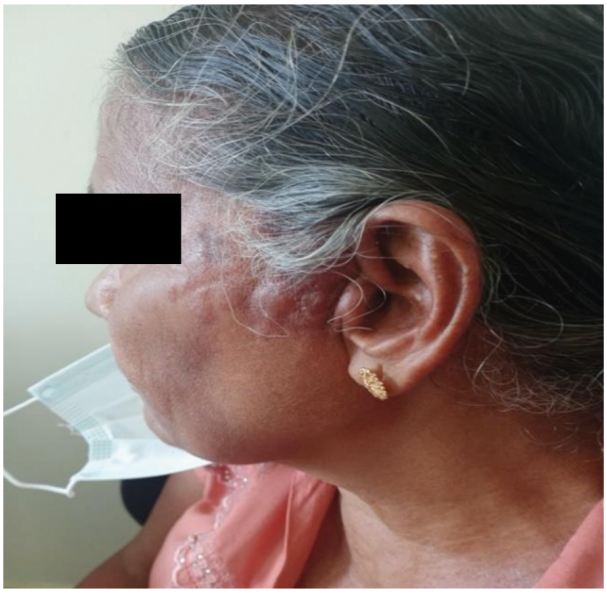

Figure 1.

Case presentation

A 70-year-old female presented with erythematous nodules on the left side of the face over the pre auricular area for 1-year duration. It gradually increased in size and was painful. It was not associated with loss of appetite, loss of weight, headache or low-grade fever. She had no other illnesses or no past surgeries.

On examination, there were well-defined erythematous multiple firm tender nodules, mainly in the pre auricular area. There was no lymph node enlargement (Figure 1). Presumptive diagnoses were granulomatous diseases, vascular neoplasm and secondary deposits. On investigations, the full blood count, ESR, CRP and liver and renal biochemical investigations were unremarkable. The skull x-ray was normal. The punch biopsy confirmed a vascular neoplasm and the deep incisional biopsy showed predominant vaso formation. Cells showed enlarged hyperchromatic nuclei and eccentric eosinophilic cytoplasm, frequent mitosis, and stroma showed collagen dissection by neoplastic cells (Figure 2).

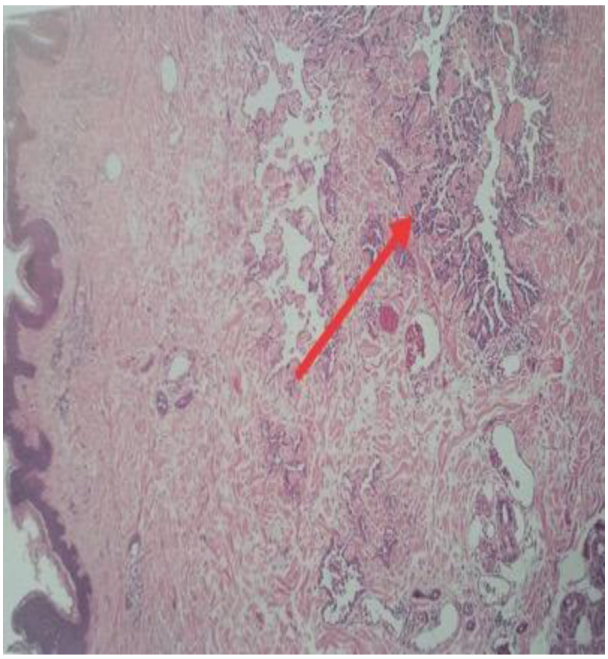

Figure 2.

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Immunohistochemical staining results

CD 31 – neoplastic cell shows strong cell membrane staining

CD 34 – neoplastic cells are negative

Pan Ck, Melanin A, HMB 45, S 100, EMA, CD 138, Desmin negative

Ki 67 proliferation index 40%

MRI of L/auricular region showed T 2 hyper intense signals (Figure 3).

CT scan of the liver and abdomen was normal.

As there was no dissemination at the time of presentation patient underwent wide complete resection of the tumour.

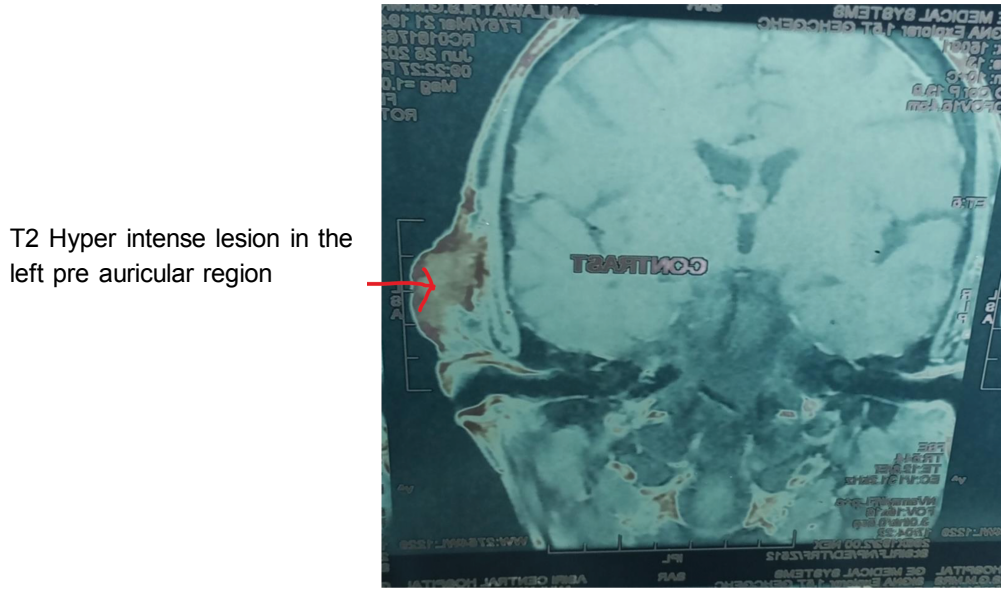

Figure 3.

Discussion

Cutaneous angiosarcoma is a rare, malignant tumour of mesenchymal origin. It first appears as an area of bruising followed by dusky blue /red nodules. Tumour disseminates early. First visceral deposits occur in the lung/pleural cavity followed by lymph node, bone and liver. Fortunately, our patient did not have visceral dissemination at the time of presentation. In the well differentiated tumour, vascular channels infiltrate the normal structure in a disorganized fashion. The collagen is lined by tumour cells in a pattern described as "dissection of collagen". Less well differentiated tumour shows more atypical pleomorphic cells. Advanced malignancy is associated with loss of vascular structures and proliferation of cell masses[6].

Immunohistochemical studies indicated that antibodies to CD 31 are the most reliable marker compared to antibodies to factor v111 and CD 34. Other histological differential diagnosis includes Kaposi sarcoma, which has eosinophilic cytoplasmic globules, melanoma which is positive for s100 and HMB 45, and lymphoma which is LCA positive.

Wide local excision with or without radiotherapy is the treatment of choice for resectable tumours. Chemotherapeutic agents like doxorubicin and paclitaxel are used for unresectable tumours and metastatic disease. Recently targeted therapy – e.g., bevacizumab (VEGF Ab), ipilimumab (antibody against cytotoxic T lymphocyte associated antigen 4 CTLA-4), nivolumab (an antibody against program me death 1 (pD 1 recepter) had been tried in some patients[7].

All angiosarcomas have a bad prognosis. Tumour size and completeness of the excision appear to be more reliable factors in predicting the outcome.

Conclusion

Based on the above history, examination, and investigation findings, the diagnosis of cutaneous angiosarcoma is made which is a very rare tumour.

References

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