

## **Aggressive epithelioid angiosarcoma of the scalp: A diagnostic challenge**

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### **Abstract**

We report the case of an 80 year old lady with multiple scalp swellings of 4 months duration. On further evaluation, the computerized tomography (CT) scan showed multiple well defined isodense to hyperdense soft tissue swellings on the left frontal region without any intracranial extension. A biopsy of the lesion was taken which on histopathological and immunohistochemical examination confirmed the diagnosis of epithelioid angiosarcoma. The patient was treated with adjuvant radiotherapy and on follow up has local recurrences at the same site six months after initial diagnosis. To conclude; epithelioid angiosarcoma is an aggressive tumour of elderly patients with local recurrences and poor prognosis.

**Keywords:** Elderly female, epithelioid angiosarcoma, scalp.

### **1. Introduction**

Cutaneous epithelioid angiosarcoma is a rare malignant tumour of vascular origin, usually located in the scalp. Epithelioid angiosarcomas are mimickers of a number of poorly differentiated tumours including carcinomas, lymphomas and melanomas.

### **2. Case Report**

An 80 year old lady presented with multiple rapidly growing swellings on the scalp of 4 months duration. She was previously treated elsewhere for multiple scalp abscesses with antibiotics. There was no past history of radiation therapy in this patient. The lady was poorly built and nourished; otherwise the general examination was unremarkable. On examination, there were multiple painless, firm to hard nodules with ulceration and surface incrustation involving the scalp and forehead. The largest nodule measured 5x4cm [Figure 1].

**Figure 1: Multiple lobated swellings on scalp showing ulceration and crusting (4X)**

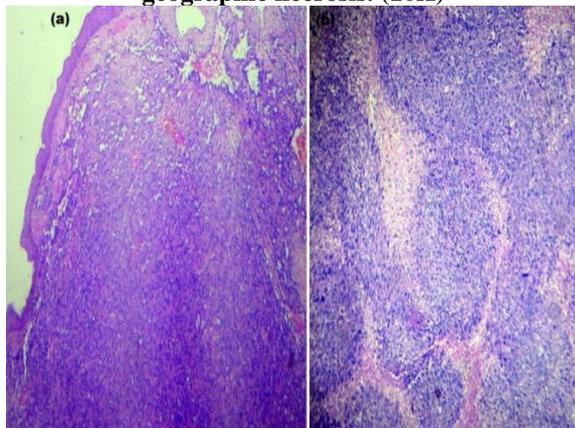


A computerized tomography (CT) scan of the head showed multiple well defined isodense to hyperdense soft tissue swellings of varying sizes over the left frontal region without any intracranial extension. Ultra sonogram of the abdomen was normal.

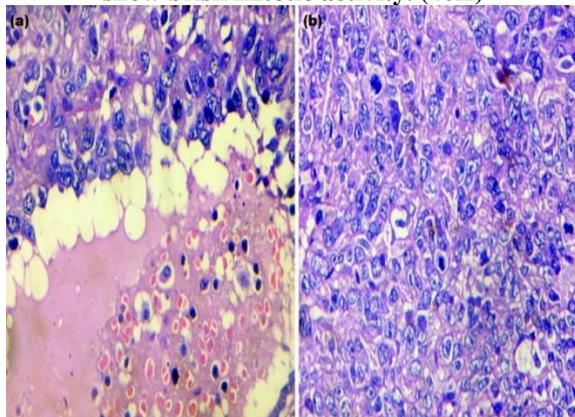
Histopathology revealed a tumour characterized by sheets of large polygonal cells with copious cytoplasm and central or eccentrically placed vesicular nuclei with prominent nucleoli. Intracytoplasmic vacuoles containing red blood cells were present. Mitosis was brisk and there were areas

displaying geographic necrosis. The better differentiated areas showed irregular anastomosing vascular channels with multi-layered enlarged endothelial cells dissecting collagen bundles [Figures 2,3,4].

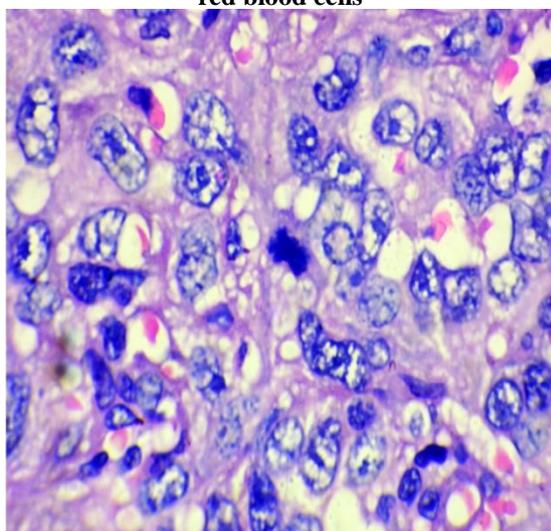
**Figure 2: (a) Well differentiated areas of the tumour showing anastomosing vascular channels (4X)**  
**(b) Islands of tumour cells with intervening geographic necrosis. (10X)**



**Figure 3: (a) Multilayered enlarged endothelial cells. (40X)**  
**(b) The cells have an epithelioid morphology and show brisk mitotic activity. (40X)**

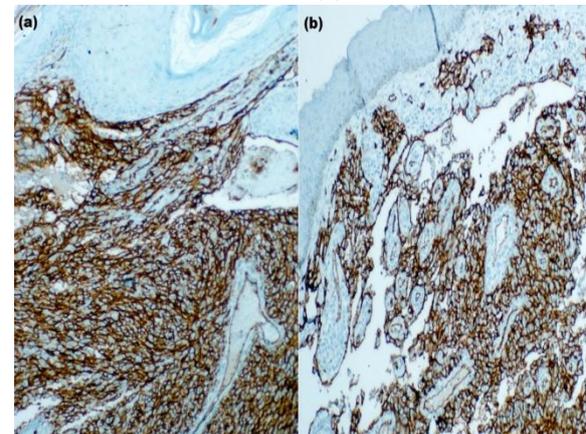


**Figure 4: Intracytoplasmic vacuoles containing red blood cells**



Reticulin staining demonstrated cellular organization into primitive vessels. Immunohistochemistry showed positivity for CD31 and CD34 [Figure 5]. The patient was treated with adjuvant radiotherapy following which surgery was planned. Clinical follow up of the patient revealed local recurrences at the same site 6 months after initial diagnosis.

**Figure 5: Immunoreactivity of tumour cells for (a) CD31 and (b) CD34**



### 3. Discussion

Angiosarcomas are rare tumours of endothelial origin which arise in various sites such as the skin, soft tissue and viscera.[2] Epithelioid angiosarcoma is an aggressive variant of angiosarcoma; usually involving soft tissues in which the malignant endothelial cells have an epithelioid appearance.[3] Cutaneous epithelioid angiosarcoma is very rare and has been addressed in only few studies. Studies show that epithelioid angiosarcoma presents in the scalp as an incrustated, raised erythematous nodular lesion; which is similar to our case.[4] The microscopic features in our case are comparable to previous studies of cutaneous epithelioid angiosarcoma. A few variations in histopathology are reported in literature such as a diffuse growth pattern with a brisk lymphocytic infiltrate resembling lymphoepithelial like carcinoma.[5] Other histological patterns include a syncytial growth pattern which may resemble any of the poorly differentiated tumours, including melanoma, lymphoma or carcinoma.[4] Another case showed plasmacytoid polygonal cells resembling a melanoma or rhabdoid tumour.[6]

The histology of this case suggests mixed origin from blood and lymphatic endothelia as there are foci in which the vascular spaces resemble lymphatic channels. There are studies which support that angiosarcomas express mixed endothelial phenotypes of blood and lymphatic capillaries.[7] This can be confirmed by positive expression of D-240. Many studies have demonstrated the presence of

Wiebel Palade bodies in few of the cells on electron microscopy.[2] Since epithelioid angiosarcoma is a mimicker of numerous non vascular tumours including carcinoma, melanoma and lymphoma, a proper clinicopathological correlation with emphasis on morphological details is essential before interpreting immunohistochemistry in difficult cases.[8] Cutaneous epithelioid angiosarcomas are associated with an unfavourable clinical outcome, repeated local recurrences and tumour related deaths.[9] Treatment modalities vary among individual cases; but surgical resection followed by adjuvant radiation therapy is usually followed.[10] There are also reports of tumour remission after combined use of adjuvant radiation therapy and bevacizumab, followed by surgery.[10]

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