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Primary Cutaneous Angiosarcoma - Scalp

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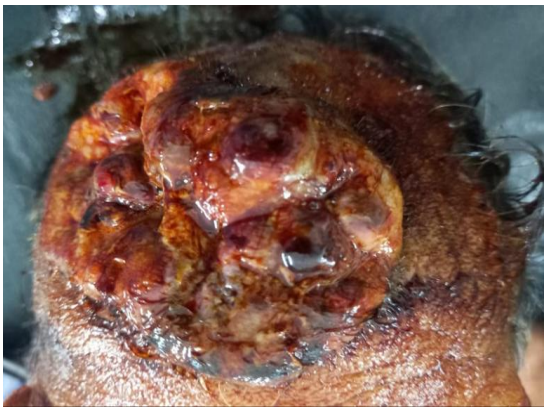
Abstract: ***This case report presents a comprehensive assessment of an 84-year-old female with an ulcero-proliferative scalp lesion, initially suspected to be either Squamous Cell Carcinoma or Amelanotic Melanoma. Radiological investigations revealed extra cranial soft tissue lesions. Histopathological examination following wedge biopsy disclosed features consistent with Primary Cutaneous Angiosarcoma, including a proliferating neoplasm of the dermis characterized by irregular anastomosing vascular channels, solid areas containing spindle cells arranged in fascicles, focal myxoid changes in the stroma, large epithelioid cells with ovoid vesicular nuclei, extensive haemorrhage, and necrosis. Immunohistochemical analysis demonstrated positivity for CD31 and Vimentin, supporting the diagnosis. This case underscores the diagnostic challenges associated with cutaneous lesions in elderly patients and highlights the importance of a multidisciplinary approach integrating clinical, radiological, and histopathological assessments for accurate diagnosis and optimal management.***

Keywords: Angiosarcoma; Cutaneous angiosarcoma; Scalp; CD31 Antigen; Vimentin

1. Case Report

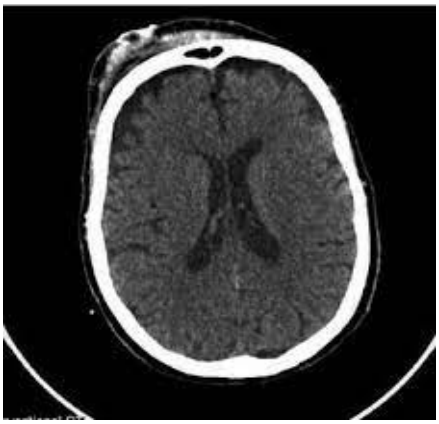
History:
An 84-year-old female presented to outpatient department with complaints of Headache for past 6 months; Swelling over Scalp for 3 months which was insidiously noticed as an ill-defined patch and later rapidly progressed to an ulcero-proliferative lesion

On Examination:
An Ulcero-proliferative lesion - 8x5 cm over frontoparietal region - multi nodular, spongy, bleeds on touch with a satellite lesion - an Ovoid violaceous lesion 4x4 cm over the occiput



Clinical Diagnosis:
Amelanotic Melanoma/ Squamous Cell Carcinoma

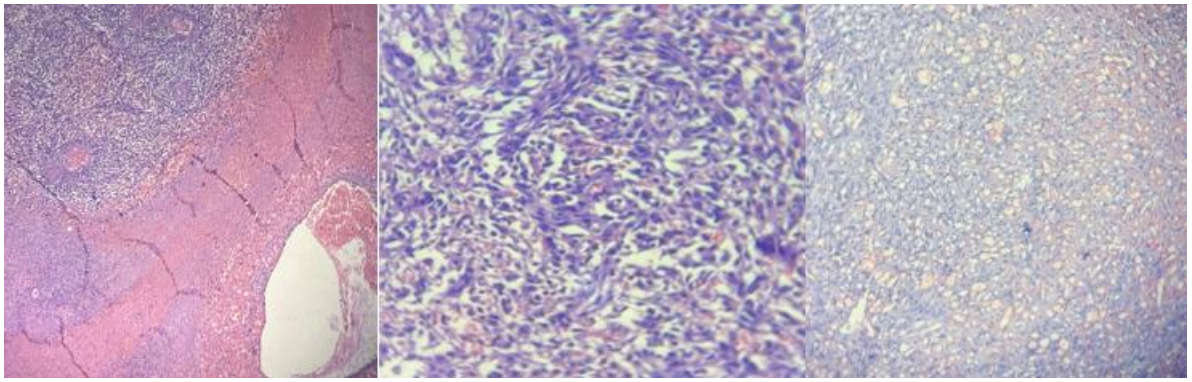
Investigations:
CT Brain: Extra cranial soft tissue lesions



Wedge Biopsy:
Hyperplastic stratified squamous epithelium with keratinous material & foreign body giant cell reaction – possibility of vascular neoplasm cannot be excluded

Treatment:
Wide local Excision with 1 cm margin till periosteum of the scalp

Histopathological Examination: Consistent with Malignant vascular neoplasm - **Cutaneous Angiosarcoma**
Histopathology description:
Proliferating neoplasm of dermis with irregular anastomosing vascular channels; Solid areas with Spindle cells as fascicles; Stroma – focal myxoid changes; Large epithelioid cells with ovoid vesicular nuclei; Extensive haemorrhage with necrosis; All Margins free of tumour; IHC: CD31+ & Vimentin +



2. Discussion

- Rare & highly aggressive malignant tumor of vascular endothelial origin
- < 2% of all sarcomas
- Adults and elderly patients; Male to Female ratio– 3:1
- 60 % - Cutaneous Lesions – Head & Neck; Retroperitoneum; Visceral organs; Bones
- Etiology: Idiopathic; Post- radiation; Longstanding Lymphedema
- 4 variants of cutaneous Angiosarcoma: Idiopathic AS (Head & Neck) - 50-60%; Lymphedema associated Angiosarcoma; Radiation induced Angiosarcoma; Epithelioid Angiosarcoma
- Post mastectomy – chronic lymphedema – Stewart Treeves Syndrome
- **Treatment of Choice:** Surgical excision with wide margins
- Whenever feasible complete resection is done despite multifocality
- Post operative adjuvant Radiotherapy – to enhance local control.
- Paclitaxel based Chemotherapy
- Targeted Immunotherapy: Anti- PDL1 antibody; Pembrolizumab; Ipilumab; Nivolumab
- Metastatic disease – Chemotherapy + Radical Radiotherapy
- Prognosis – Very poor prognosis - Median survival of 2 years

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