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Case Report

Angiosarcoma- Scalp- Multimodal Approach

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ABSTRACT

Angiosarcoma is a rare and highly aggressive malignant tumour of vascular endothelial origin, accounting for less than 1% of all soft tissue sarcomas (1). It most commonly affects the head and neck region, particularly the scalp in elderly individuals (2). Management depends on tumour extent and typically involves wide surgical excision with appropriate reconstruction (3). Adjuvant radiotherapy and chemotherapy are considered in cases of locally advanced, recurrent, or metastatic disease (4). We report the case of a 73-year-old woman presenting with multiple rapidly progressive scalp lesions associated with fungation and necrosis. Imaging revealed a large subgaleal lesion with calvarial erosion but no intracranial extension. The patient underwent wide local excision followed by reconstruction using a transposition flap, split-thickness skin grafting, and vacuum-assisted closure therapy. Following satisfactory wound healing, adjuvant chemotherapy was initiated. This case highlights the aggressive nature of scalp angiosarcoma and the importance of multimodal management.

Keywords: *Angiosarcoma, Scalp Neoplasms, Vascular Tumors, Surgical Excision and Reconstruction, Multimodal Therapy.*

INTRODUCTION

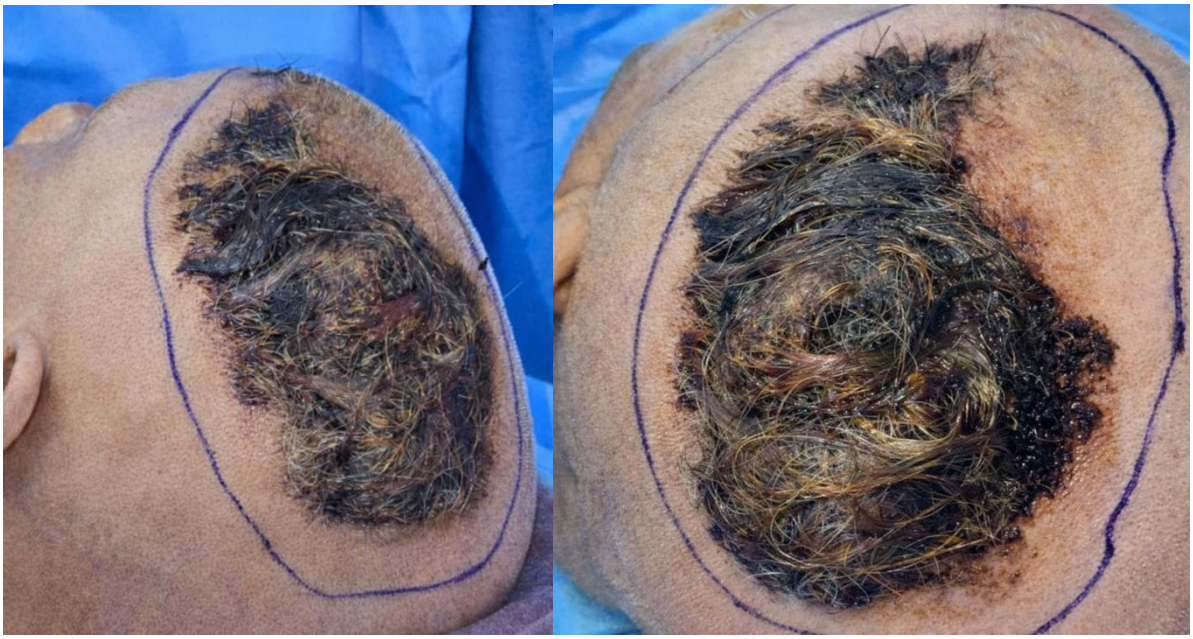
Angiosarcoma is a rare malignant neoplasm arising from vascular endothelial cells, characterized by aggressive biological behavior and a high propensity for local recurrence and distant metastasis (1,5). The etiopathogenesis of angiosarcoma remains multifactorial. Established risk factors include prior radiation exposure, chronic lymphoedema (Stewart–Treves syndrome), and environmental carcinogens such as arsenic and vinyl chloride (8,9). Histopathologically, angiosarcomas demonstrate a wide spectrum ranging from well-differentiated vasoformative lesions to poorly differentiated tumours composed of highly atypical endothelial cells with marked mitotic activity (5,10). It most frequently involves the skin and superficial soft tissues of the head and neck region, particularly the scalp and face in elderly individuals (2,6). Clinically, lesions may initially resemble benign conditions such as bruises or ecchymoses, often leading to delayed diagnosis (7).

Given its infiltrative growth pattern and tendency for early dissemination, management remains challenging. Surgical excision with negative margins remains the cornerstone of treatment, often supplemented by systemic chemotherapy and radiotherapy in advanced disease (3,4). Despite aggressive therapy, prognosis remains poor, with reported 5-year survival rates ranging from 10% to 35% (6,11).

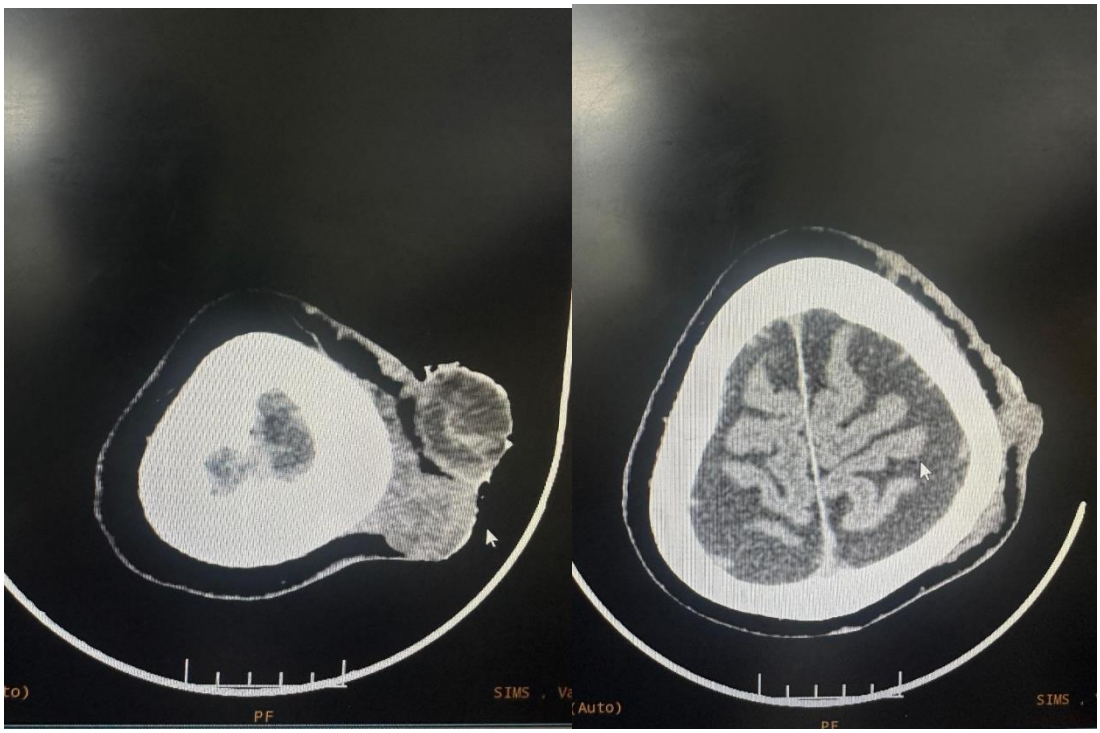
CASE REPORT

A 73-year-old woman presented with multiple lesions over the frontoparietal region of the scalp, which had progressively increased in size over a short duration. The lesions were associated with serosanguinous discharge and foul odor.

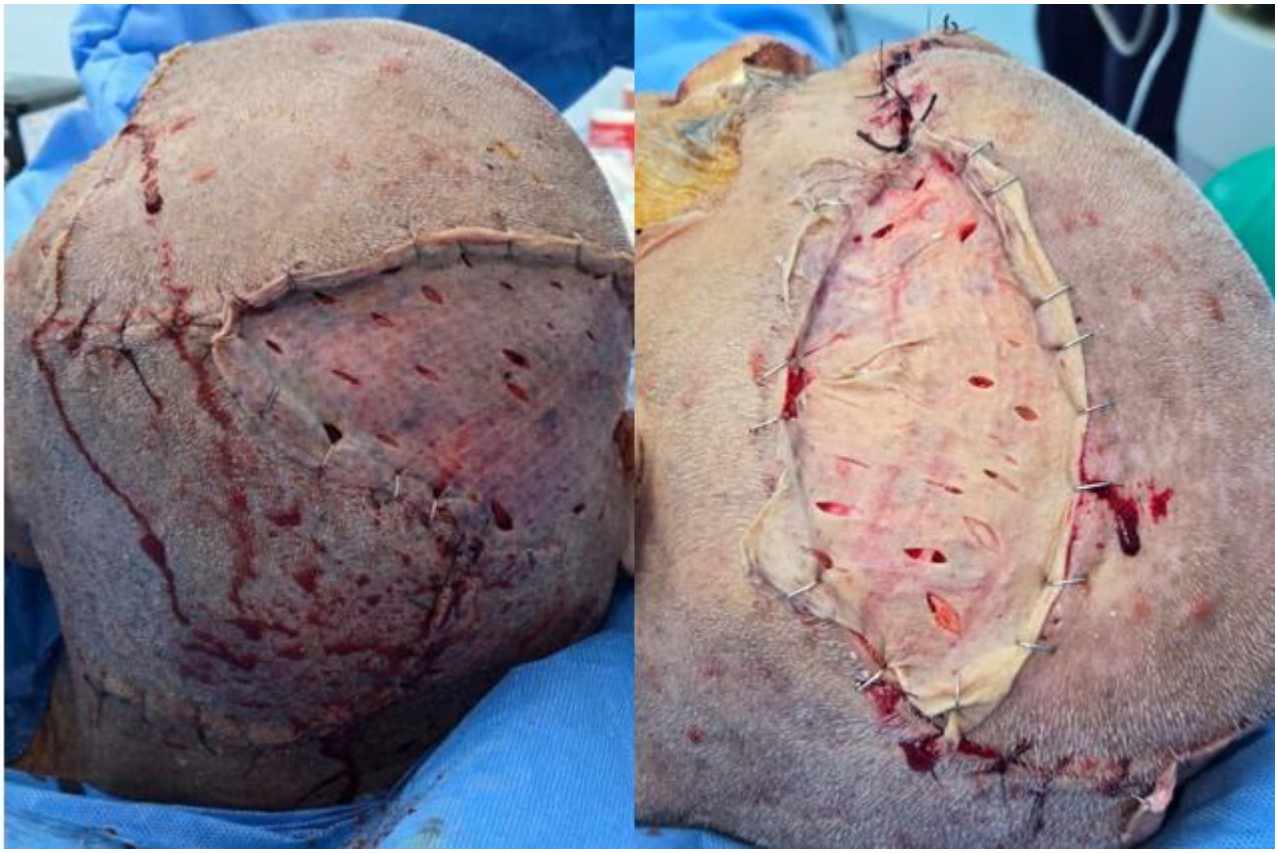
On clinical examination, a large irregular mass measuring approximately 20 × 10 cm was noted over the left parietal region. The lesion appeared brownish-black with areas of ulceration, necrosis, and fungation, and had a nodular consistency. Evidence of subcutaneous spread with multiple satellite nodules over the frontal and parietal scalp was present.



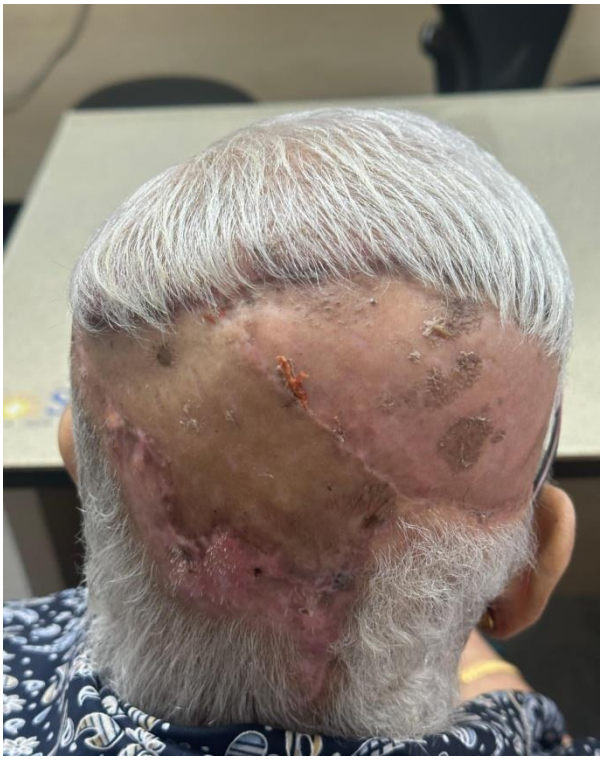
Contrast-enhanced computed tomography of the head demonstrated an ill-defined, bilobulated, heterogeneous hypodense soft tissue lesion in the subgaleal plane with an exophytic component. There was associated nodular thickening of the adjacent scalp and erosion of the underlying calvarium, without intracranial extension.



An incision biopsy revealed a poorly differentiated malignant neoplasm suggestive of angiosarcoma. The patient subsequently underwent wide local excision of the tumour with adequate margins. Reconstruction was performed using a transposition flap mobilized from the occipital region to cover the primary defect. A split-thickness skin graft harvested from the left thigh was applied over the donor site and areas with exposed calvarial bone. Vacuum-assisted closure therapy was done to promote granulation tissue formation.



The postoperative course was uneventful, with flap and graft take up very well. After complete wound healing, medical oncologist opinion was obtained and advised VAC regimen including Vincristine, Adriamycin and Cyclophoshamide for 3 cycles. The patient is doing well, on regular follow-up, and is planned for reassessment after three cycles of the VAC regimen before initiating radiotherapy.



DISCUSSION

Angiosarcoma represents a biologically aggressive subset of soft tissue sarcomas, with marked heterogeneity in histological differentiation (5,10). Well-differentiated tumours are characterized by irregular, anastomosing vascular channels lined by atypical endothelial cells, whereas poorly differentiated variants exhibit solid sheets of pleomorphic cells with high mitotic activity, necrosis, and hemorrhage (10).

Cutaneous angiosarcoma of the scalp predominantly affects elderly males, although it can occur in females as demonstrated in the present case (6). The typical presentation includes bruise-like patches that may progress to nodules, ulceration, and bleeding lesions (7). Advanced lesions often exhibit fungation and extensive local invasion, reflecting delayed diagnosis. Angiosarcoma has one of the highest rates of lymphatic and hematogenous spread among head and neck sarcomas (11). The lungs are the most frequent site of distant metastasis, followed by the liver and bones (6,11). Recurrence is common even after complete surgical excision, necessitating long-term surveillance (3).

Surgical excision with histologically negative margins remains the primary treatment modality. However, achieving clear margins in scalp lesions can be challenging due to the infiltrative and multicentric nature of the tumour (3,6). Reconstruction depends on defect size and depth, with options including skin grafts, local flaps, and free tissue transfer. Vacuum-assisted closure therapy serves as a valuable adjunct in managing complex wounds with exposed bone.

Adjuvant radiotherapy has been shown to improve local control, particularly in patients with close or positive margins (4). Chemotherapy, particularly taxane-based regimens such as paclitaxel, and anthracyclines, has demonstrated efficacy in advanced or metastatic disease (12). Emerging therapies, including targeted agents and immunotherapy, are currently under investigation.

CONCLUSION

Scalp angiosarcoma is an aggressive malignancy with a high risk of recurrence and poor overall survival. Early diagnosis and prompt, aggressive multimodal treatment are essential to improve outcomes. Wide surgical excision with tumour-free margins remains the cornerstone of management, while adjuvant therapies play a crucial role in advanced disease. Given the risk of delayed recurrence and distant metastasis, lifelong surveillance is imperative (3,11).

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