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CASE REPORT

Angiosarcoma of the scalp in a 79-year-old male: A case report

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ABSTRACT

INTRODUCTION Angiosarcoma is a rare head and neck sarcoma of vascular endothelial cell origin. We report a case of angiosarcoma in an elderly male, and the multidisciplinary approach employed in his treatment.

CASE REPORT A 79-year-old male presented with a 4-month history of a rapidly enlarging black, soft, immovable tumor surrounded by bruise-like patches over the right temporoparietal scalp. There was associated pruritus and bleeding when scratched. Dermoscopy showed bluish black crusts over the tumor, and surrounding violaceous patches. Wedge biopsy revealed a dermis with irregular vascular spaces infiltrating dermis, lined by atypical endothelial cells. Immunohistochemistry of the atypical infiltrative cells was positive for CD31. These findings were consistent with angiosarcoma. The patient underwent wide excision with a rotational flap and split thickness skin graft. Postoperatively, the patient was referred to Oncology for adjuvant radiation therapy.

CONCLUSION Even with treatment, the prognosis of angiosarcoma remains poor due to its aggressive nature, with a 5-year survival rate ranging from 10-54%. However, early detection of the disease may increase patient survival rates. This rare case shows the importance of maintaining a high level of suspicion for lesions that have an atypical presentation to prevent delays in management and improve patient outcomes.

KEYWORDS Angiosarcoma, scalp, oncology, neoplasm

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INTRODUCTION

Angiosarcoma is a rare, malignant tumor of vascular endothelial cell origin.[1,2] It has an incidence of approximately 0.01/100,000 per year, and comprises <2% of soft tissue sarcomas and 5% of malignant skin tumors.[1,3,4] It can occur in any site, although the most common locations for the primary tumor are the skin of the head and neck region.[3-5] There are 3 clinical variants – primary cutaneous angiosarcoma, lymphangiosarcoma or Stewart-Treves syndrome, and post-irradiation angiosarcoma.[1,3,6]

Primary cutaneous angiosarcoma is difficult to diagnose clinically due to its variable presentation. The lesion may present as singular or multifocal, appearing as bruise-like patches, plaques or nodules that may bleed or ulcerate.[1,7] Prognosis is poor, and treatment is difficult due to the aggressive nature of the neoplasia, and a lack of standardized protocol for the management of the disease.[3,6,7]

We report a case of angiosarcoma in an elderly male, and the multidisciplinary approach employed for his treatment.

CASE REPORT

This is a case of a 79-year old male presenting with a 4-month history of a rapidly enlarging brown to black plaque surrounded by bruise-like patches over his right temporoparietal scalp. There was associated pruritus and bleeding when scratched. He was a known hypertensive, maintained on amlodipine and losartan. The patient initially sought consult with General Surgery, and ultrasound was requested; the results showed a cutaneous growth with increased vascularity confined to the basal region. A diagnosis of basal cell carcinoma was considered. The patient was then referred to the Dermatology service for further evaluation.

On cutaneous examination, there was a solitary, well-defined, black, soft, immovable tumor with a violaceous base measuring 3.5 x 2.8 x 2.1 cm over the right temporoparietal scalp (Figure 1). Dermoscopy findings were nonspecific, showing bluish black and hemorrhagic crusts. There were

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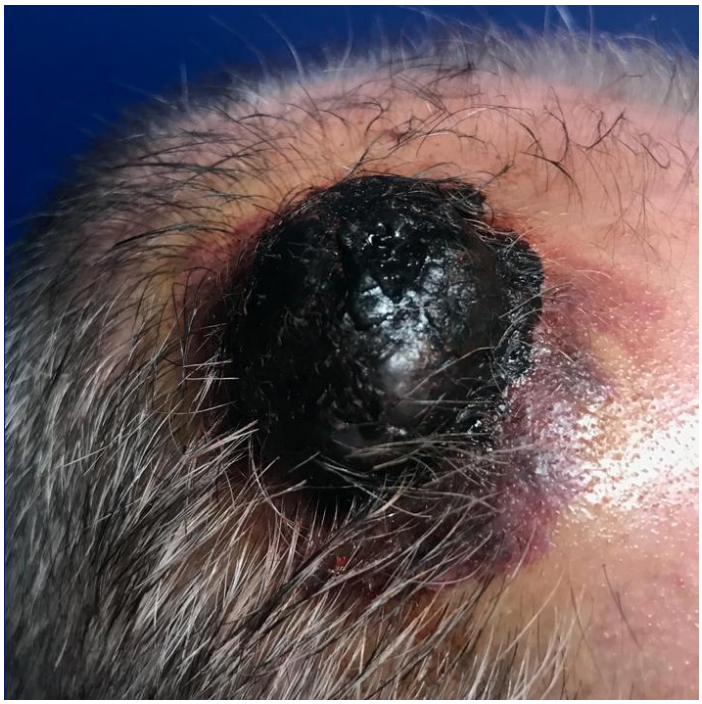

Figure 1. Solitary, well-defined, black, soft, immovable tumor with a violaceous base over the right temporoparietal scalp.

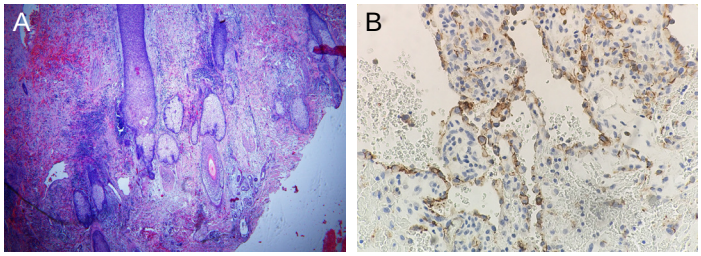

Figure 2. **A.** Hematoxylin and eosin stain revealed irregular vascular spaces infiltrating the dermis, lined by atypical endothelial cells. **B.** Immunohistochemistry revealed atypical infiltrative cells positive for CD31.

no palpable periparotid, perifacial, buccinator, and cervical lymphadenopathies.

A wedge biopsy of the tumor was done, and histopathology revealed irregular vascular spaces infiltrating the dermis, lined by atypical endothelial cells (Figure 2a). Immunohistochemistry of the atypical infiltrative cells was positive for CD31 (Figure 2b). These findings were consistent with a diagnosis of Angiosarcoma.

To evaluate the extent of the tumor, a cranial computed tomography (CT) scan was done. The imaging revealed a 3.7 x 3.1 x 5.3 cm, well-defined, lobulated, heterogenous mass on the right parietal extra-calvarial region, exhibiting minimal heterogenous enhancement with no associated calvarial erosions and intracranial metastasis. Contrast enhanced chest CT scan was also done, which revealed multiple, non-calcified pulmonary nodules, suspicious for pulmonary metastasis.

The patient was referred back to General Surgery – Head and Neck service for surgical evaluation and management. Palliative wide excision of the scalp lesion was done. A 2 cm margin from the violaceous base was marked using Gentian violet prior to surgical incision. The mass resected was hard and slightly movable, measuring approximately 5 x 4 cm. Frozen section examination of resection margins showed tumor deposits at the basal margins. The periosteum was also removed as an additional basal margin, which eventually revealed negative for tumor deposits. The Plastics and Reconstructive Surgery service closed the surgical defect using full thickness scalp rotational flap with split thickness skin grafting. A flap was developed from the medial aspect of the defect and the anterior scalp. The flap was then rotated to cover the defect. A superficial partial thickness skin graft was harvested from the left thigh, and was placed into a 1:1.5 mesher. The skin graft was then placed on the defect on the anterior scalp (Figure 3).

Microscopic section showed a malignant tumor constituted by epithelioid or fusiform cells with rudimentary vascular differentiation. Pleomorphism, mitosis, and tissue infiltration were present. Surgical margins of resection were devoid of tumor involvement. This case was signed out as Scalp Angiosarcoma stage IV (T2N0M1).

The patient was compliant with follow up at the outpatient department, with note of viable flap and skin graft on subsequent weeks postoperatively. The patient was referred to the Oncology service for co management. He underwent 25 cycles of radiation therapy. He was started on palliative chemotherapy with Paclitaxel, though he was only able to complete 2 cycles. Eight months post-surgery, the patient was admitted due to dyspnea. He eventually expired due to acute respiratory failure secondary to poor ventilatory drive.

DISCUSSION

Angiosarcoma is difficult to diagnose and treat, not only because of its aggressive nature, but also due to its rarity. Most published studies are case reports and retrospective analyses. There are no clinical or randomized trials to standardize diagnostic and therapeutic management.[2,4]

Clinical diagnosis is difficult due to the variable presentation of angiosarcoma. Presenting signs may be ill-defined, bruise-like macules or patches that progress to more indurated plaques or nodules. Most lesions are painless, though there may be bleeding and ulceration.[6,7] Similarly, our patient presented with a painless, brown to black plaque that rapidly progressed to a tumor with a violaceous base, and a tendency to bleed. Misdiagnosis on first consult is fairly common due to the nonspecific clinical picture, and the low suspicion for the disease.[5]

Definitive diagnosis of angiosarcoma can be made through

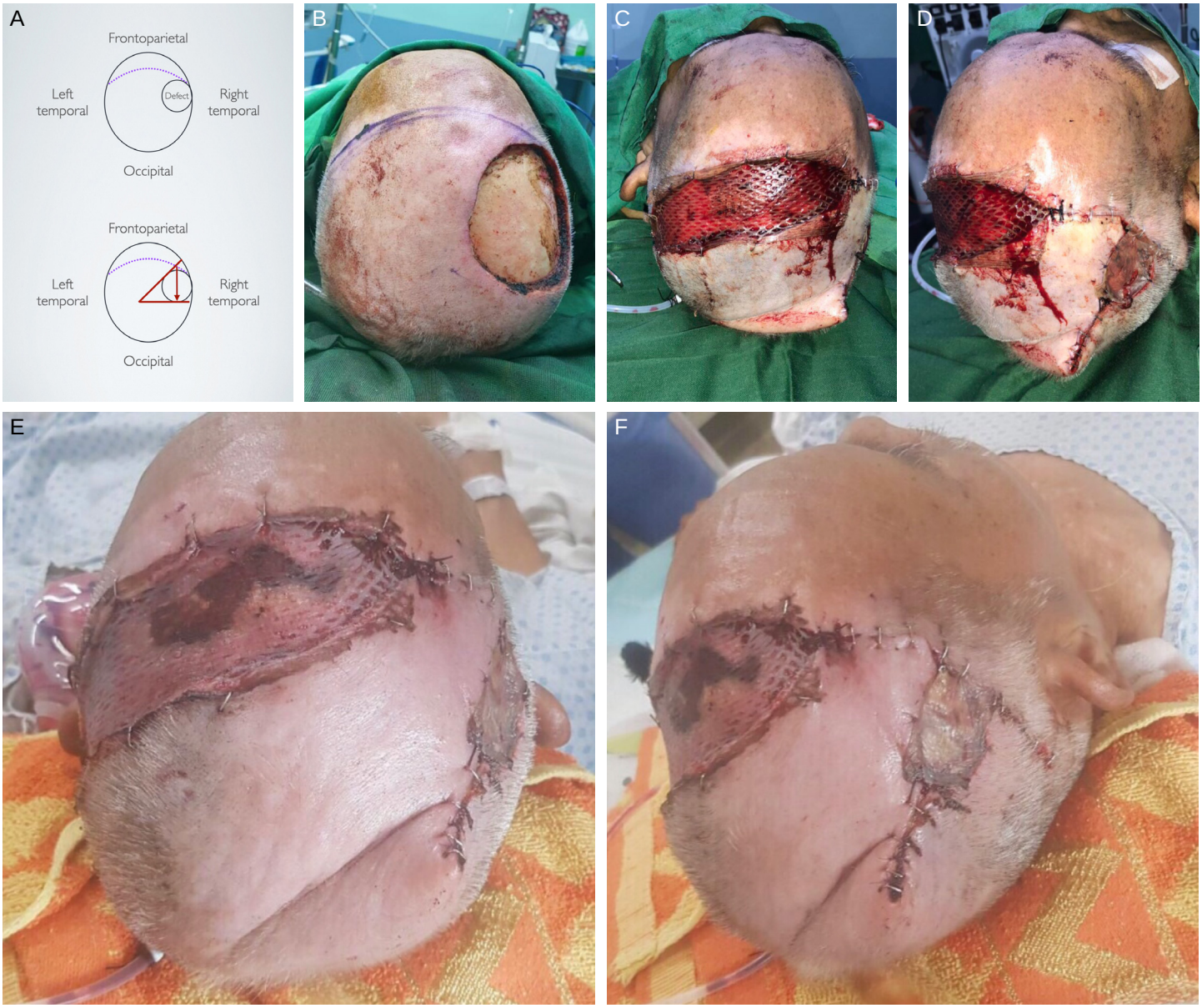

Figure 3. **A.** Diagram of the procedure **B-D.** intraoperative images of the procedure. **E-F.** Day 5 postoperative images of the surgical site.

histopathologic examination.[5] Angiosarcoma extensively involve the dermis, with a possible invasion of deeper structures such as fascia, and subcutaneous tissue.[7] For this reason, a wedge biopsy may be the better technique for collecting sample as it can provide the dermatopathologist a deeper view of the specimen. A punch biopsy may provide inadequate tissue, and an excision biopsy may be difficult due to the tumor's location and the challenge of achieving negative margins.

Angiosarcoma is characterized by irregular, anastomosing vascular channels lined by endothelial cells that dissect through collagen.[1,6,8] Nuclear atypia is always present. Mitotic figures are variable, though a high mitotic rate may be associated with a poor prognosis.[6,8] These histopathologic findings were consistent with our patient's biopsy.

Specific markers for angiosarcoma include factor VIII antigen, CD34, and CD31. Of the three, CD31 is the most sensitive and specific endothelial marker that stains positive in more than 90% of angiosarcomas.6 In this case, the specimen stained positive for CD31, confirming a diagnosis of angiosarcoma.

The rarity of the disease and its clinical variability

may contribute to a delay in diagnosis, which averages to 5.1 months.[5,7] Due to this delay, multiple lesions, extensive local growth, or metastasis may already occur prior to detection of the disease.[7] The most frequent sites of metastases include the lung, and lymph nodes.[3,7] Though the patient was initially asymptomatic, pulmonary metastasis was considered due to the multiple nodules seen in his chest CT scan. Another challenge encountered was the delay in complete diagnostic work-up due to financial constraints.

The current treatment of choice remains to be wide local excision even though it is difficult to achieve negative margins due to its multifocal growth tendency.[1,3,4,7] There are no standardized recommendations as to what constitutes as adequate margins, though a margin <1 cm is associated with poor survival.[10] Mohs micrographic surgery was not considered due to the tumor's tendency for multifocal growth, the difficulty of achieving negative margins,[4] and pulmonary metastases. Our patient's cranial CT scan revealed that the mass was extra-calvarial, with no calvarial erosions. Given the advanced age of the patient and the suspicious findings of possible distant metastases, the surgical treatment included a more conservative wide local excision, ensuring negative pathologic margins to include only the full thickness of the scalp layer and not the skull bone. This was to ensure prevention of early local recurrence of the tumor on the surgical defect, as more radical resection will not give any further benefit to the patient in terms of overall survival.

Post-operatively, the patient was referred to Oncology for adjuvant radiation therapy and chemotherapy. Studies show that this may improve local control and survival rates.[2,4,7] Subsequent follow-up should be done every 3-6 months for the first 2-3 years, then every 6-12 months thereafter. Complete history and physical examination must be done to assess for recurrence.[9]

The prognosis for patients with scalp angiosarcoma is poor, with a 5-year survival rate ranging from 10%-54%.[2,3,7] Factors associated with poor outcomes include tumors > 2 in or 5 cm, patients >70 years old at presentation, deep location of the tumor, and presence of necrosis.[3,4,7] Our patient presented 2 of these factors, specifically the tumor size, and advanced age.

CONCLUSION

Angiosarcoma is a rare, malignant tumor that poses a diagnostic challenge for clinicians. It is important to maintain a high index of suspicion for lesions that have an atypical presentation in order to prevent delays in diagnosis. The sarcoma may present as ill-defined, bruise-like macules or patches that progress to more indurated plaques or nodules with bleeding and ulceration. Early diagnosis could be associated with a smaller tumor size and absence of metastasis on presentation of the disease, possibly increasing the survival rate for patients.[3] Though limited, current observations show the benefit of a multidisciplinary approach, including dermatopathology, surgery, and oncology in the management of patients with angiosarcoma.

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