Letter to the Editor

Primary Angiosarcoma Masquerading as Scalp Haematoma After Head Injury

Dear Editor,

Angiosarcomas are extremely uncommon, making up <1% of all sarcomas.[1] Specifically, the scalp accounts for approximately 50% of all angiosarcoma cases, yet this comprises a mere 0.1% of all head and neck malignancies.[2] Delayed diagnosis often occurs due to difficulty in recognising this rare tumour.[3] Presumably, due to the location preference of this neoplasm, it has been previously shown that minor head trauma may sometimes alert the patient to the presence of a lesion.[4] We report a case of an elderly male who presented with what was presumed to be a scalp haematoma after a minor head injury. Given the rarity of this condition, the case is discussed in corroboration with current literature and management strategies.

Case Report

A previously well 76-year-old male presented to the neurosurgery outpatient clinic with a painless right parietal scalp lump. The patient described a history of persistent scalp swelling after minor trauma to his head approximately a month earlier. He had knocked his head against the edge of an overhead cupboard while at work. There had been some bleeding at the time of accident but this had resolved spontaneously. At the beginning, he visited his local polyclinic a few times. The patient was informed that he had an infected scalp haematoma. A trial of oral antibiotics and local dressings were prescribed but the swelling did not resolve. A decision was then made by his primary care physician to refer him to a specialist.

Clinical examination demonstrated a single, well circumscribed 3 cm raised scalp lesion that was mildly tender and centrally fluctuant. The nodular lesion had shiny and indurated edges. The edges also showed an underlying bluish tinge under close examination. Hair was observed on the dark-coloured surface of the lesion. There was no active discharge or bleeding from the lesion. There were no constitutional symptoms and the rest of the patient's system review was otherwise unremarkable. A postcontrast computed tomography (CT) scan of the head reported a non-enhancing, soft tissue mass at the high right parietal scalp region measuring about 2.8 × 2.1 cm. There was no significant surrounding fat stranding. The deep aspect of the lesion was noted to abut the underlying parietal bone but there was no radiological evidence of bony erosion or remodelling. The Hounsfield unit of the scalp lesion was 44 on both pre- and postcontrast CT images. This confirmed that the lesion had no or minimal contrast uptake (Fig. 1).

Decision was made for an incision biopsy of the lesion (Fig. 2). Histopathology showed a tumour consisting of pleomorphic spindle and epithelioid cells with mitotic activity. Immunohistochemical stain was positive for CD31, FLI-1 and ERG; and negative for AE1/3, p40, S100 and HMB45. The final report was that of angiosarcoma. As a result of the diagnosis, the patient underwent extensive staging investigations. The investigations demonstrated no radiological evidence of metastases.

The patient underwent en-bloc resection of the lesion with a generous 4 cm margin and an outer table craniectomy underlying the lesion. Intraoperative frozen section biopsies taken around the resection margins showed these to be tumour-free. An omental free flap was harvested laparoscopically and was used to cover the resultant 25 × 14 cm scalp defect. This flap was successfully anastomosed with the superficial temporal artery and vein. A split-thickness skin graft was

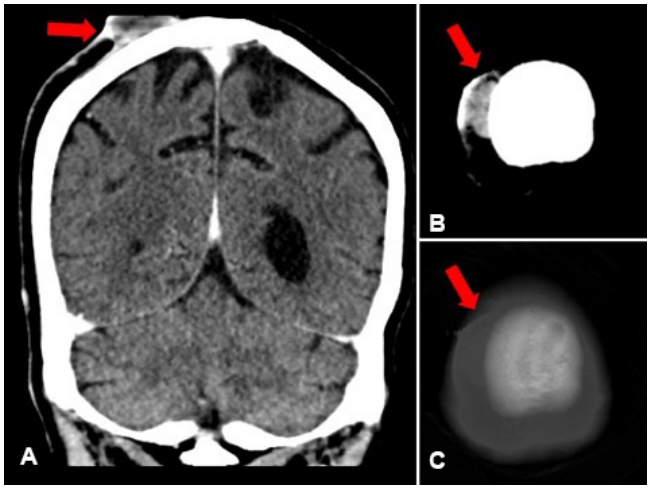

Fig. 1. Computed tomography (CT) images of the brain. A and B: Postcontrast images of the extracranial, soft-tissue mass in right high parietal scalp region in coronal (A) and axial (B) views, respectively. C: Axial bone window view that corresponds to B shows no radiological evidence of bony involvement under the extracranial, soft-tissue scalp mass.

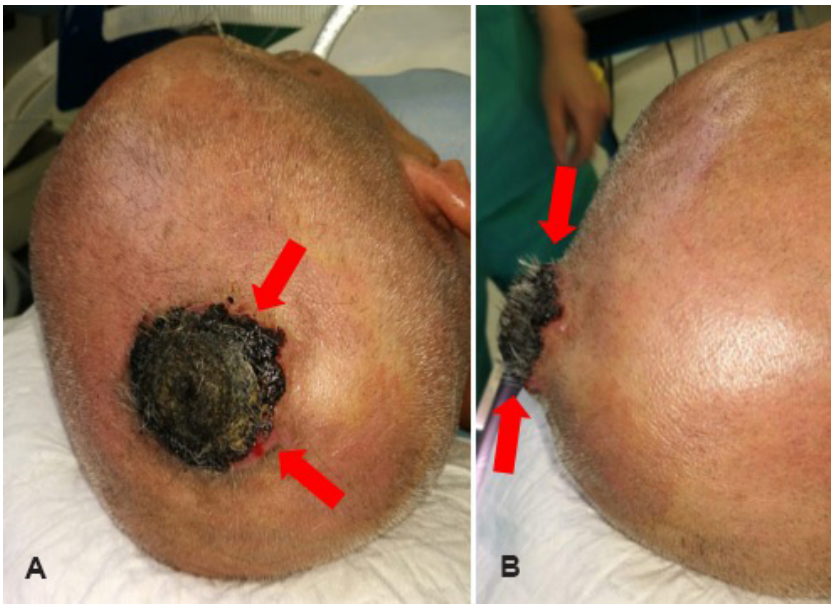

Fig. 2. Intraoperative photographs of the right parietal scalp mass showing the top (A) and side (B) views. The patient's scalp had been shaved. A broad-based and raised soft-tissue lesion with hair on its surface can be seen. There is evidence of induration at the scalp-lesion interface. No active bleeding was noted from the lesion.

harvested from the thigh and subsequently grafted onto the omental flap. Final histopathology concurred with the previous diagnosis of angiosarcoma (Fig. 3). In addition, the surgical margins were reported to be tumour-free.

Postoperatively, the patient's recovery was uneventful. Approximately 2 months after his surgery, he commenced radiation therapy. However, 1 month into his treatment, the patient developed type 1 respiratory failure. An urgent CT thorax reported multiple lung nodules within the parenchyma of both lungs. Some of the nodules were cavitating in nature. There was also bilateral hydropneumothoraces associated with pleural nodularity. Putting it all together, the working diagnosis was lung metastases. Attempts were made at draining the associated malignant pleural effusions but the fluid kept re-accumulating. Decision was made for commencement of chemotherapy once the patient was clinically more stable. However, the patient continued to deteriorate rapidly and eventually succumbed about 18 weeks following his en-bloc excision surgery.

Discussion

Angiosarcoma is an infrequent but aggressive disease that portends a poor prognosis. The aggressive lesions arise from vascular endothelial cells and tend to grow along pre-existing vascular channels, sinusoidal or cavernous spaces; often forming poorly organised vessels, solid masses or nodules.[5]

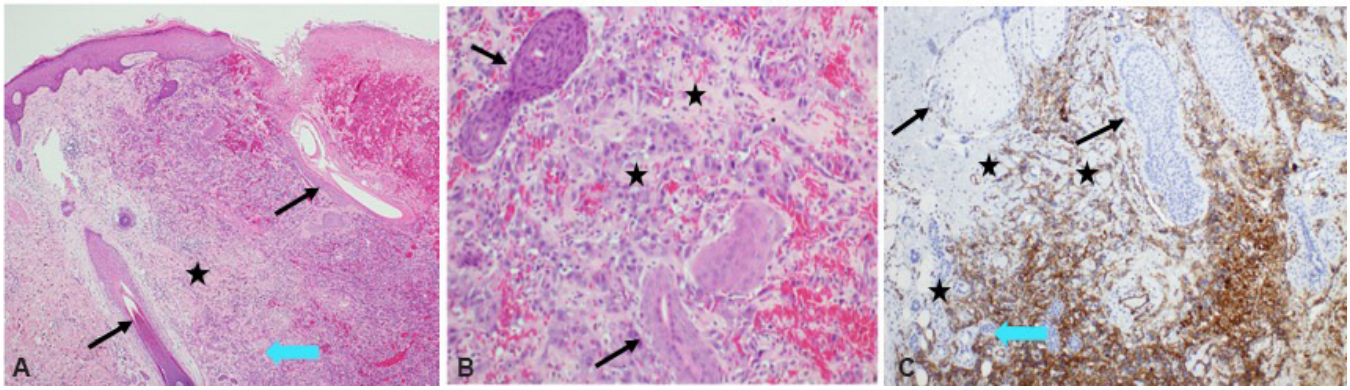

Fig. 3. A: Low-power view of the skin of the scalp reveals a cellular tumour associated with ulceration and haemorrhage. The tumour infiltrated around the skin adnexal structures, which were hair follicles (black arrows) and sweat glands (blue arrows) (haematoxylin and eosin [H & E] stain, × 40). B: The tumour consisted of pleomorphic spindle and epithelioid cells forming vascular structures and dissecting between dermal collagen bundles. Asterisks denote representative dermal collagen fibres where the tumour was dissecting between them (H & E stain, × 200). C: Immunohistochemical stain of the vascular marker CD31 showed tumour cells infiltrating around skin adnexal structures and dissecting between dermal collagen bundles (H & E stain, × 100).

Men have a higher risk of this disease than women (ratio, 1.7:1).[6] Established risk factors for this tumour include previous exposure to chemical toxins (such as vinyl chloride and arsenic), chronic lymphoedema,[7] radiation exposure and certain rare genetic syndromes.[1] Common differential mimics—which may lead to delayed diagnosis—include skin ecchymoses, capillary haemangioma, rosacea, eczema, cellulitis and angioedema.[2,3,6] The condition has also been reported to be only noticed after what was thought to be a minor skin injury of the scalp.[2,4] Patients may present with skin lesions such as a bruise, discoloured nodule or persistent ulceration.[2] Another reason for the initial difficulty in early diagnosis is that primary cancers of the scalp are extremely rare.[8] They can present as ecchymosis-like plaques, associated with haemorrhage, oedema and ulceration.[9] Amongst the imaging modalities for malignant skin neoplasms, CT is the most readily available and least costly.[10] It is able to delineate tumour extent and staging. On contrast-enhanced CT, soft tissue angiosarcoma may manifest as an irregular, enhancing soft tissue mass. In more advanced cases, underlying bone or adjacent solid organ invasion may be present.[2,11] However, this was not the case for our patient; hence, getting the correct initial diagnosis was challenging.

An estimated 20–30% of affected patients present with metastatic disease, and about 50% of patients with primary tumours will develop early interval metastases.[6] The usual sites for metastases—in decreasing order of incidence—include the lungs, bone and liver.[6,12] As a rule of thumb, radical surgery aims to reduce tumour burden. Nonetheless, surgery is unable to target "subclinical" micro-metastases, which could have occurred by the time of diagnosis.[13] These lesions may be too small to be picked up by conventional imaging techniques. Generally, angiosarcoma has a poor overall survival range of between 6–16 months.[1,2] Despite best efforts, the majority of patients succumb to disease recurrence or metastases. For our patient, it was unusual that he developed metastases close to the completion of his radiotherapy. However, given the aggressive nature of his primary tumour, it is not unexpected that such an event is likely to occur.

In addition, histopathological factors associated with worsening prognosis that include the presence of epitheloid morphology and necrosis[6,14,15] were unfortunately observed in our patient. These factors have been demonstrated in clinical studies focusing on stratification markers in cutaneous angiosarcoma.[14,16] However, there is currently no effective targeted therapy for these tumours.

The current standard of care for patients with localised angiosarcoma is wide surgical resection with histopathologically clear margins, followed by adjuvant radiotherapy.[6,17,18] This may be technically challenging as the tumour tends to be widely infiltrative beneath the skin leading to difficulty in obtaining negative margins.[1] Although there are no clear guidelines on the specific width of the surgical margins in localised angiosarcoma, widths of ≥3 cm have been recommended for both radial and deep margins.[6] Generally speaking, if the tumour is localised and operative margins are free of malignant cells, the disease can be considered curative. Conversely, if the patient has recurrent and/or metastatic disease, treatment is difficult as there remains a lack of effective chemotherapeutic and/or targeted therapy. At present, we are aware that primary angiosarcomas are genetically complex. They are reputed to have gene mutations such as *TP53*, *KRAS PTPRB* and *PLCG1* involving various oncogenic pathways.[19] To complicate matters in this rare tumour, studies have shown that primary and secondary angiosarcomas possess different molecular profiles.[6] In comparison to other cancers, the delineation of biological subtypes for angiosarcomas is still unclear at this stage. Owing to current limited adjuvant options, there is thus a global need for urgent preclinical research to elucidate the pathogenesis of this deadly tumour. Radical excision of the tumour with clear margins remains as the mainstay of curative treatment.

Conclusion

We have, in this article, looked into a case of a primary scalp angiosarcoma in a 76-year-old male who presented with what was presumed to be a scalp haematoma secondary to minor head trauma. This case strongly emphasises the need for clinicians to consider angiosarcoma as a differential diagnosis when encountering scalp lesions in the elderly, especially in the primary care setting. Early intervention is a key priority in managing this malignant disease. In addition, we advocate for more in-depth molecular studies. For patients, we recommend that they attain a better understanding and insight of the disease.

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