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Case Report

Primary angiosarcoma of the skull: A rare case report

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Abstract

Background: Angiosarcomas are rare high grade endothelial tumors characterized by rapidly proliferating anaplastic cells derived from blood vessels and lining irregular blood filled spaces. Primary neoplasms of the skull are rare, representing 2.6% of primary neoplasms of bone. Primary malignant neoplasms of the skull are even rarer, accounting for only 0.8% of primary malignant neoplasms of bone.

Case Description: We report a 32-year-old female who presented with right parieto-occipital swelling, which gradually increased in size. Radiology was suggestive of a calvarial soft tissue lesion in the right parieto-occipital region with destruction of the adjacent parieto-occipital bone with intracranial extra-axial extension. Complete surgical excision of the calvarial lesion was done under general anesthesia. Postoperative computed tomography (CT) scan of brain (plain and with contrast) showed complete excision of the tumor mass. Histopathological diagnosis was consistent with 'an angiosarcoma of the skull'. On immunohistochemistry, the atypical endothelial cells were highlighted by CD34, CD31, and factor VIII-related antigen. The patient received adjuvant radiotherapy to the tumor bed.

Conclusion: Primary angiosarcoma of the skull is a rare tumor with less than 20 cases reported worldwide till date. The treatment should include complete surgical excision with a wide bony margin followed by adjuvant radiotherapy, which in our case has given a good locoregional control even at the end of 2 years. However, these patients should be followed up with repeated scans yearly to rule out locoregional as well as distant recurrence.

Key Words: Angiosarcoma, female, parieto-occipital, skull

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INTRODUCTION

Angiosarcomas are high grade aggressive neoplasms, which may occur in any region of the body but occur more frequently in the skin and soft tissues of the body. Angiosarcomas may also originate in the liver, breast, spleen, bone, or heart.[1,21,23,27]

Angiosarcoma of the skull is a rare entity with less than 20 cases reported in literature.[26] It may present either as a primary tumor or as a metastasis of a distant extra-cranial angiosarcoma. Angiosarcomas have a poor prognosis and the prognosis is worst if the location is the skull. This is because of the frequent secondary involvement of the brain and the meninges and the

difficulty to perform a complete surgical resection in some cases.

CASE REPORT

A 32-year-old female was admitted to our institution with a 5-month history of swelling in the right parieto-occipital region . The swelling gradually increased to the present size of about 8 × 8 cm. The skin overlying the swelling appeared normal and the scalp was mobile over the swelling. There was no relevant past history such as head injury. Her family history was unremarkable. Her blood profile for biochemistry and hematology was within normal limits. The neurological examination was unremarkable except for exaggerated deep tendon reflexes on the left side of the body and left sided extensor plantar response (positive Babinski sign). There was no regional lymphadenopathy. A detailed examination of the whole body was performed to rule out any cutaneous or subcutaneous lesions.

Computed tomography (CT) scan of brain (plain and with contrast) and magnetic resonance imaging (MRI) scan of brain (plain and with contrast) were done. Findings were suggestive of a calvarial soft tissue lesion in the right parieto-occipital region with destruction of the adjacent parieto-occipital bone with intracranial extra-axial extension. The lesion appeared hyperintense on spin-spin relaxation time (T2)/fluid attenuated inversion recovery (FLAIR) images. It also showed peripheral hyperintense rim on spin-lattice relaxation time (T1) images with postcontrast enhancement, not suppressed on fat suppression or fat saturation (FAT SAT) images. Dimensions of the lesion were 5.4 × 4.3 × 5.4 cm. There was indentation of the sulco-gyral spaces but no midline shift was seen. There was indentation of the posterior aspect of the superior sagittal sinus at places [Figure 1]. On MRI, the differential diagnosis of the lesion was epidermoid or dermoid cyst. Additional studies like contrast enhanced computed tomography (CECT) thorax, abdomen, and pelvis and bone scan were negative for any metastatic lesions.

Patient was taken for surgery and complete excision of the calvarial lesion was done under general anesthesia in prone position [Figure 2]. A linear incision was taken over the swelling. The galea appeared normal and so we could raise a plane between the galea and the tumor. The tumor was in the form of a complex cyst with intratumoral hemorrhage. A wide craniectomy up to the normal bone was done. The tumor was easily dissected from the underlying duramater. Except for some hyperemia, the duramater appeared normal and so it was not resected. Throughout the operation, an epidural plane was maintained. We did not attempt to reconstruct the bony defect at the time of initial surgery. Elective cranioplasty with Codman™ cranioplastic bone cement was done after completion of the adjuvant radiotherapy. The patient had an uneventful postoperative hospital stay and did not show signs of any neurological deficit. Postoperative CT scan of brain (plain and with contrast) showed complete excision of the calvarial lesion [Figure 1]. On her latest follow up, 2 years postsurgery, there is no locoregional or distant recurrence.

Pathology

Microscopic sections of the tumor showed dilated irregular vascular spaces filled with blood and fibrin. These spaces were lined by pleomorphic atypical endothelial cells. Many giant and bizarre tumor cells were also seen. Large areas of hemorrhage were seen. On immunohistochemistry, the atypical endothelial cells were highlighted by CD34, CD31, and factor VIII-related antigen. These findings were consistent with angiosarcoma of the skull [Figure 3].

DISCUSSION

Angiosarcoma is a malignant neoplasm of the vascular endothelium. It rarely involves the bone. About 50% of cases of primary bone angiosarcoma involve the long bones of the extremities.[3,25] Less frequently, the ribs, pelvis, or vertebrae may be involved. Primary skull angiosarcoma is a rare presentation with less than 20 cases reported in literature so far.[26] Radiation therapy, arsenic exposure, and history of previous trauma are known risk factors for extra-cranial angiosarcomas. On the contrary, primary skull angiosarcoma has no known risk factors.[5,16]

Primary skull angiosarcoma has a 2:1 male predominance, with a median age of 32 years.[16] The most common presentation of a skull angiosarcoma is a swelling in the affected region. Additional clinical manifestations like

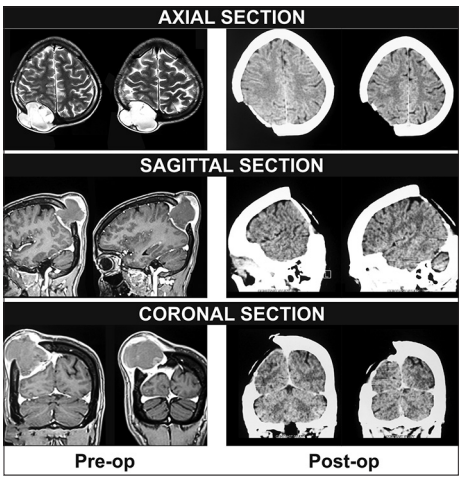

Figure 1: Pre-operative MRI of brain (plain and with contrast) showing a calvarial soft tissue lesion in the right parieto-occipital region with destruction of the adjacent parieto-occipital bone with intracranial extra-axial extension. Post-operative CT scan of brain (plain and with contrast) showing complete excision of the calvarial lesion

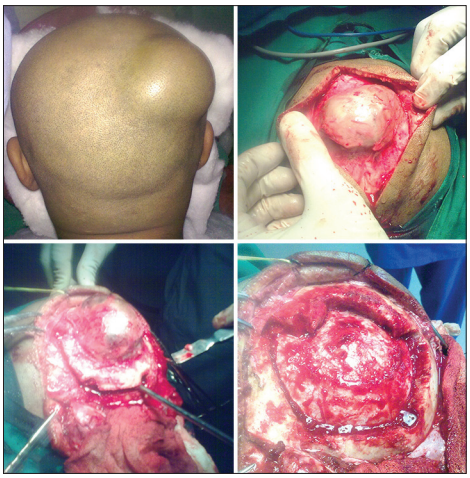

Figure 2: Complete surgical excision of angiosarcoma of the skull in process

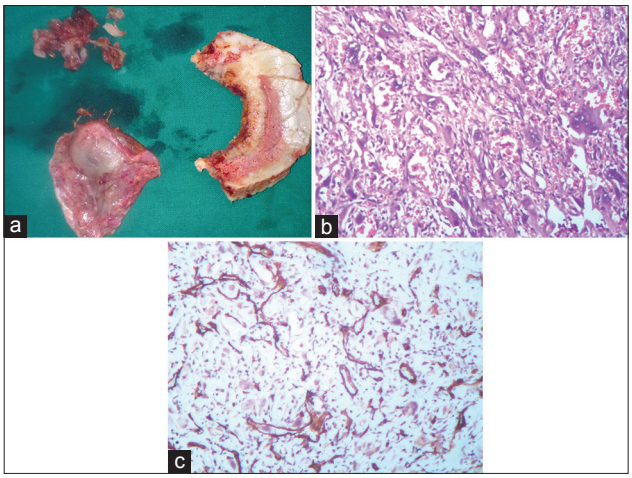

Figure 3: (a) Gross specimen of angiosarcoma of the skull. (b) Microscopic picture which on H and E shows highly vascular tumor. There are many dilated irregular vascular spaces lined by pleomorphic atypical endothelial cells.(c) Immunohistochemistry (CD34): The atypical endothelial cells are highlighted by CD34

hearing impairment, tinnitus, and otalgia may be present with a swelling in the temporal region in temporal bone angiosarcoma.[20] Shuangshoti *et al.* reported a case of angiosarcoma involving the lesser and greater wings of the sphenoid bone, frontal bone, and roof of the orbit causing unilateral proptosis.[22] Khan *et al.* described a case of primary calvarial angiosarcoma who presented with a single episode of left limb numbness and weakness due to brain parenchymal compression.[12] Yamada *et al.* reported a case of epithelioid angiosarcoma causing subdural hematoma.[26]

A plain skull X-ray usually demonstrates a well demarcated, expansile, lytic lesion.[10,24] In the skull, the frontal bone is the most commonly involved. MRI brain (plain and with contrast) usually shows a mass that is isointense with gray matter on T1-weighted images and hyperintense on T2-weighted images.[2]

The differential diagnosis of primary skull angiosarcoma includes meningioma, hemangiopericytoma, primary intravascular papillary endothelial hyperplasia, epithelioid sarcoma, and metastasis.[22] In poorly differentiated tumors, immunohistochemistry is mandatory for identification of an endothelial lineage. The markers of endothelial lineage are factor VIII-related antigen, CD31, and CD34, which are positive in cases of angiosarcoma, as seen in our case.

Patients with primary skull angiosarcoma usually have a poor prognosis. This is because of the frequent secondary involvement of the brain and the meninges and the difficulty to perform a complete surgical resection in some cases. Also, these tumors metastasize early due to invasion of the rich intradiploic blood supply of the skull.[3] Metastasis usually occurs to the bone and lungs.[6] According to Mark *et al.*, in a report of 67 patients with angiosarcoma, 52 patients developed recurrences after primary treatment. Of these 52, 42 patients had a component of local failure and 28 patients had developed distant metastases at last follow-up.[17] Because of the strong metastatic potential, an extensive staging work up should be performed in all cases at the time of diagnosis. According to Bourekas *et al.*, the follow up should include MRI of the region of surgery for evaluation of recurrence as well as bone scan and chest radiography for evaluation of metastatic disease.[2]

Because of the rarity of skull angiosarcomas, optimal management has not been defined. After going through the literature, the most effective treatment is complete surgical excision of the tumor followed by adjuvant radiotherapy.[14,17] Since angiosarcomas are highly vascular tumors, preoperative selective arterial embolization using materials such as foam and springs may be performed, 24 h before surgery, to reduce the amount of blood loss during surgery.

Radiotherapy has been used as an adjunct to surgical treatment or as palliative treatment.[5,9,11,13] Chow *et al.* reported a case of an angiosarcoma of the frontal bone, which was treated with neoadjuvant radiotherapy (6000 cGy over a 6-week period) followed by complete surgical excision of the lesion with a 2 cm margin of grossly uninvolved bone around the lesion. On histopathological examination of the resected specimen, only fibrosis and suture granuloma were seen. No residual tumor was identified.[6] Scholsem *et al.* and Khan *et al.* have also reported the use of adjuvant radiotherapy with good local control at the tumor site.[12,20]

Recent data suggest that adjuvant chemotherapy with Paclitaxel has shown good results in patients with soft tissue sarcoma of the face or scalp.[4,7] However, in the case reported by Scholsem *et al.*, the patient received adjuvant chemotherapy consisting of 6 cycles of Paclitaxel at the dose of 175 mg/m^2 but it could not prevent the development of lung and bone metastasis.[20]

Recently, Bevacizumab has been used in the adjuvant setting for the treatment of cutaneous facial angiosarcomas with good results.[8,15,18,19] Whether the same holds true for primary skull angiosarcoma needs to be investigated.

CONCLUSION

Primary angiosarcoma of the skull is a rare tumor with less than 20 cases reported worldwide till date.[26] The treatment should include complete surgical excision with a wide bony margin followed by adjuvant radiotherapy, which in our case has given a good locoregional control even at the end of 2 years. However, these patients should be followed up with repeated scans yearly to rule out locoregional as well as distant recurrence.

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