

Case Report

Spindle cell sarcoma of sphenoid bone

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ABSTRACT

Primary bone tumors involving skull are extremely rare and they constitute 0.8% of all bone tumors. The common tumors that are seen in skull base include fibrous dysplasia, giant cell tumor, chordoma, ossifying fibroma, angiosarcoma. We report a rare case of spindle cell sarcoma arising from right sphenoid bone in a 70-year-old male which presented as unilateral defective vision with mild proptosis.

Keywords: MR imaging, Sphenoid bone, Spindle cell sarcoma

INTRODUCTION

Sphenoid bone is a butterfly shaped bone at the base of skull. The parts of the sphenoid bone include body, a pair of greater wings and lesser wings and two pterygoid processes. It encloses the sphenoid sinus. Besides optic foramina, foramen rotundum, foramen spinosum and foramen ovale are seen in the sphenoid bone. Primary bone tumors involving sphenoid bone are rarely seen. The common tumors seen primarily arising from sphenoid and tumors that extend from adjacent structures include fibrous dysplasia, ossifying fibroma, chordoma, giant cell tumor, rhabdomyosarcoma, juvenile angiofibroma, neuroblastoma, angiosarcoma.¹ Both CT and MR scans are the most important imaging for preoperative evaluation of the masses.

CASE REPORT

A 70-year-old male patient presented with right sided proptosis with decreased vision since, six months. He had only perception of light on right side. There was no history of previous trauma to eye. Routine laboratory investigations were normal. CT orbits showed a 4.8 x 2.3 cms mildly hyper dense mass arising from greater wing

of sphenoid bone right side with thinned out outer cortex, cortical disruption and retro orbital extension displacing lateral rectus muscle superiorly, indenting on optic nerve and causing narrowing of superior orbital fissure. PET CT showed increased uptake in the right sphenoid bone. MRI brain including orbits with contrast enhancement showed a well-defined homogeneously enhancing lobulated mass from the greater wing of sphenoid bone on right side. The mass was hypointense to gray matter on T1 weighted sequence and hyperintense on both T2Weighted and FLAIR sequences with restriction on diffusion weighted imaging. There was no blooming on gradient sequence. Laterally the mass extends into infratemporal space and anteriorly extends into retro orbital region displacing the eyeball infero medially. There was no intracranial extension of the mass. The greater wing of sphenoid shows massive resorption. A provisional diagnosis of primary malignant bone tumor of sphenoid bone was made. Biopsy from the mass showed the evidence of low grade spindle cell sarcoma from sphenoid bone. Immuno histochemistry findings: EMA: negative, CD34: Weak positivity, Pancytokeratin-weak positivity, MIB Index– 5-10 %. Patient received external radiation therapy to right orbit and right sphenoid bone with IMRT technique 5600cGy/28 frs@200 cGy/fr.

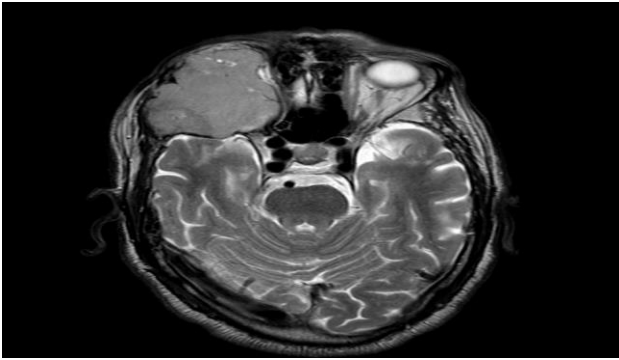


Figure 1: Axial T2weighted image of brain show hyperintense mass from greater wing of sphenoid right side.

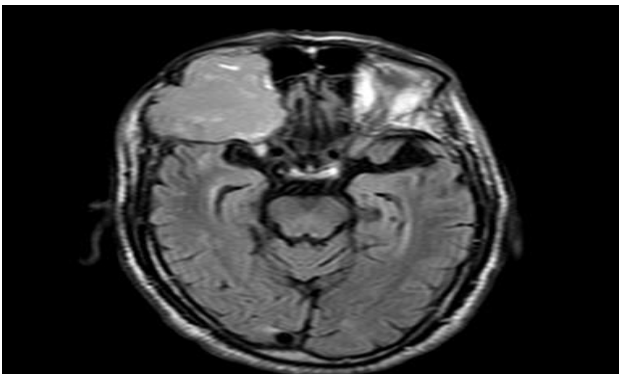


Figure 2: Axial flair image of brain show lobulated hyperintense mass.

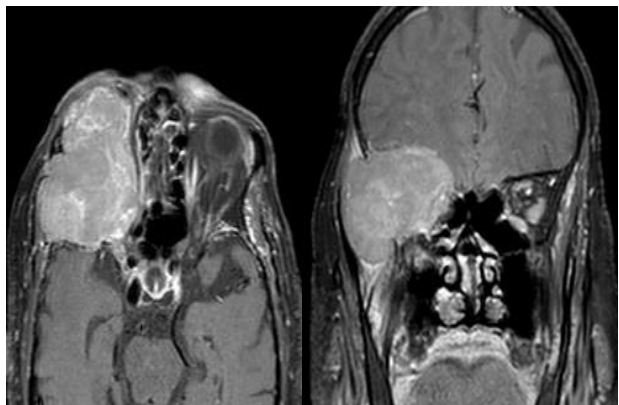


Figure 3 (A and B): Axial and coronal images brain show homogenous contrast enhancement of the mass.

DISCUSSION

Spindle cell sarcoma is a rare tumor and constitutes 2-5 % of all primary bone malignancies occurring in patients above 40 years. It arises from the connective tissues of bone, cartilage, muscle and fat. Three types of spindle cell sarcoma have been described. They are

- Pleomorphic undifferentiated sarcoma,
- Fibrosarcoma,

- Leiomyosarcoma.

The common sites of involvement are femur, tibia, fibula and pelvic bones. Spindle cell sarcoma is reported sometimes to arise from preexisting osseous disorders like giant cell tumor, osteomyelitis, bone infarction, Paget's disease, previous radiotherapy in the affected areas, non-ossifying fibroma. The common bone tumors that involve the base of skull including sphenoid bone are mostly fibrous dysplasia, giant cell tumor, angiosarcoma etc. Fibrous dysplasia: often involve cranium and facial bones. On CT scan it gives a typical ground glass appearance with dense calcification and intact cortical bone. On MRI it appears as hypo or isointense on T1 weighted images and variable signal intensity on T2 weighted images.

The lesion enhances on contrast administration. Giant cell tumor: benign bone tumor that rarely involves sphenoid bone in the cranium. On MR they are isointense to gray matter on both T1 and T2 weighted imaging. They enhance with contrast administration and seen mostly in patients below 40 years. Angiosarcoma: malignant tumor of endometrium, rarely involve sphenoid bone. Occur around 3rd decade. The other bone tumors that occasionally affect sphenoid bone include osteoma Osteoblastoma, aneurysmal bone cyst, giant cell tumor, giant cell reparative granuloma, primary Ewing' sarcoma, primary osteosarcoma, myxoid chondrosarcoma and metastases.²⁻¹²

Meningioma involving the sphenoid usually presents as sheat like mass (meningioma enplaque) and produces hyperostosis of underlying bone.^{13,14} The differential diagnosis of this condition includes all bone lesions that cause massive osteolytic destruction and soft tissue mass. The treatment protocols of spindle cell sarcoma include wide surgical excision, chemotherapy (methotrexate, cisplatin) and radiotherapy in some cases. Since spindle cell sarcoma rarely involves sphenoid bone this case has been reported.

CONCLUSION

Spindle cell sarcoma of the bone has also to be considered in the differential diagnosis of aggressive osteolytic destructive lesions of sphenoid bone.

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