CASE REPORT

# Primary Aggressive Osteosarcoma of Sphenoid Bone

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## ABSTRACT

Osteosarcoma is a malignant intra-osseous neoplasm producing osteoid. Osteosarcoma of the skull is very rare and it usually involves the cranial vault. The occurrence in the skull base is extremely rare. We report a case of primary osteosarcoma in 59-year-old lady, occur in left sphenoid wing with intracranial and intranasal extension manifesting as left facial pain, headache and left epistaxis. She underwent radiotherapy as the surgical resection of the tumour is not feasible due to the critical extent of the tumour. We describe the clinical, radiological and histopathological findings of the case.
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## INTRODUCTION

Osteosarcoma is the commonest non-haematopoietic primary malignant bone tumour accounting for 20% of cases. The metaphyseal part of the long bones is the site of predilection, particularly the distal femur (30%), the proximal humerus (15%) and the proximal tibia (15%) (rich in proliferative growth plates). The incidence of primary osteosarcoma of the craniofacial bones represent less than 10% of all osteosarcomas (1). The mandible and the maxilla are the most frequently involved bones. The incidence of primary osteosarcoma of the skull ranges between 1-2% (2). It typically involves the cranial vault and rarely involves the cranial base. To the best of our knowledge, there are only 10 cases of skull osteosarcoma that have been reported in the literature (Table I). We herein report a case of a primary osteosarcoma of the sphenoid wing in an elderly lady who presented with left facial pain and epistaxis.

## CASE REPORT

This was case of a 59-years-old lady with underlying diabetis mellitus and hypertension who presented with left facial pain for one-month duration. It was associated

**Table I: Clinical review of ten previously published primary osteosarcoma of skull (3)**

| Authors | Years | Age(y), Sex | Location | Surgery | Chemotherapy/ Radiotherapy | Follow up | Outcome |
|---|---|---|---|---|---|---|---|
| Marks et al | 1991 | 14,M | Skull base | Not Reported | One cycle/24 Gy | 12mo | Death |
| Salvati et al | 1993 | 11,M | Skull base | Subtotal resection | No/ 60Gy | 9mo | Death |
| Hayashiet al | 2000 | 28,M | Sphenoid bone | Total resection | One cycle/50 Gy | 10mo | Death |
| Chennupati et al | 2008 | 14,F | Clivus | Biopsy | One cycle/50 Gy | 12mo | Remission |
| Chou et al | 2009 | 22,F | Parietal bone | Total resection | One cycle/No | 8 y | Free of disease |
| Patibandla et a | 2011 | 30, F | Occipital bone | Near-total resection | Six cycles/53 Gy | 16mo | Dealth |
| Kirby et al | 2011 | 16,M | Parieto-occipital bone | Total resection | One cycle/No | 5mo | Free of disease |
| Meel et al | 2012 | 10,M | Sphenoid bone | Biopsy | Six cycles/50 Gy | 18mo | Free of disease |
| Mohindra et a | 2014 | 55,M | Clivus | Total resection | Six cycles/45 Gy | 1 y | Free of disease |
| Hadley et al | 2014 | 14,M | Parietal bone | Total resection | One cycle/No | 16mo | Free of disease |

with on and off headache, altered sensorium and left epistaxis. Otherwise, there was no other symptoms of increased intracranial pressure such as vomiting or blurring of vision. On examination, she had stable vital signs with Glasgow Coma Scale (GCS) of 14/15 (E4V4M6). The cranial nerves were intact. Limbs motor examination showed power of 4+/5 on left side (positive pronator drift). Sensory, tone and reflexes were normal. Cerebellar examination was normal. Other systemic examination was unremarkable. Serum alkaline phosphatase (ALP) level was high, 2000 U/L (reference range: 42 – 98 U/L). Computed Tomography (CT) brain showed an ill-defined heterogeneous hyperdense mass with calcification which arising from the left sphenoid bone. Magnetic Resonance Imaging (MRI) showed a heterogenous and ill-defined lesion with isointense signal on T1W, T2W and was not suppressed on FLAIR, seen arising from the left sphenoid bone region which extend superiorly into the middle cranial fossa and inferiorly to the left maxillary sinus, middle and inferior turbinate and posterior nasal fossa. It measured approximately 3.1cm x 4.9cm x 8.5cm (AP x W x CC). It extended into the middle cranial fossa as an extra axial lesion causing mass effect to the adjacent left temporal lobe, left lateral ventricle, left thalamus, left basal ganglia, pons and midbrain. (Fig. 1)

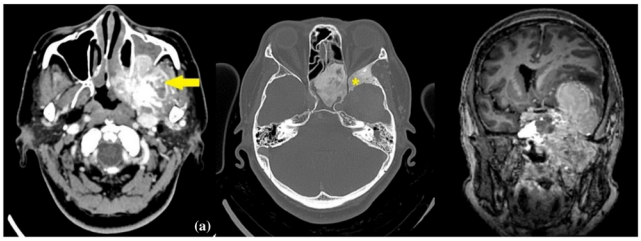


**Figure 1: (A & B) CT brain showed heterogenous enhancing lesion (arrow) arising within left sphenoid bone. Presence of area of sclerosis (*) and thickening within the sphenoid bone noted. (C) On MRI the lesion extended into the intracranial fossa causing mass effect and midline shift.**

Subsequently, patient underwent left front temporoparietal craniectomy and had an open biopsy. Intraoperatively, the tumour extended superficially at the inferior temporal gyrus up to the skull base and bled profusely upon contact. The tumour was firm in consistency and highly vascularised. The patient also underwent Examination Under Anaesthesia (EUA) through nasal cavity which revealed a mass obscuring the posterior choanae of the left nostril that bled easily and thus preceeded with left nasal packing. Whereas the right nostril was normal. Based on histopathological examination, the excised mass from choanae revealed a diagnosis of osteosarcoma. Microscopically, there were a few fragments of tumour cells intimately associated with malignant osteoid formation. The malignant osteoid formations were arranged in lace like pattern comprises of irregular eosinophilic and basophilic thin bony trabeculae. The malignant cells were composed of atypical spindle cells with hyperchromatic nuclei and minimal cytoplasm. Several osteoclast multinucleated giant cells were also noted (Fig. 2 A&B). In areas, epithelioid appearing tumour cells were appreciated characterized by large vesicular nuclei having prominent nucleoli and abundant eosinophilic cytoplasm (Fig. 2C). Mitotic figures were easily seen. These cells were positive for Special AT-rich sequence-binding protein 2 (SATB2) immunohistochemical stain (Fig. 2D).

Post operatively, the patient underwent palliative radiotherapy with tumour dose of 30 Gy given in 10 fractions of 3 Gy per day, 5 days a week for 2 weeks duration. Patient was not keen for adjuvant chemotherapy. A repeat CT scans of the brain after the completion of therapy (3 months of follow-up) did not show any significant changes in the tumour dimensions. CT scan thorax and hepatobiliary ultrasound did not show any distant metastasis. Other work - ups for primary disease were negative. The latest serum ALP level was low after three months post-operation and reported as 1657 U/L. Currently, the patient is under palliative care and bed-bound with a percutaneous endoscopic gastrostomy (PEG) tube for feeding. The symptoms are improving.

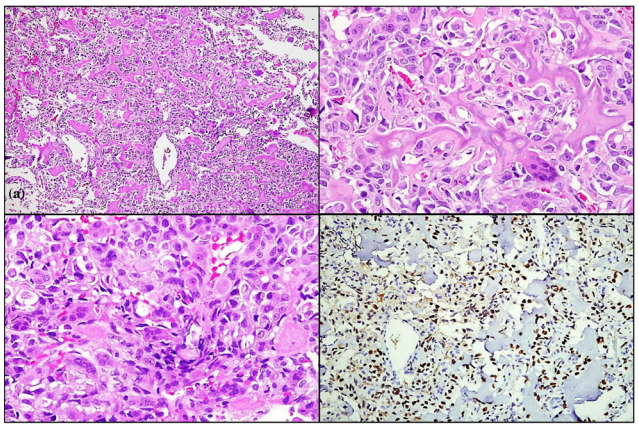


**Figure 2: (A) A low power view of osteosarcoma exhibiting a lace-like pattern of malignant osteoid (arrow) formation surrounded by atypical spindle cells (H&E x100). (B) The osteoclast type multinucleated giant cells (H&E x400). (C) Some of the tumour cells are epithelioid morphology (H&E x400). D) The tumour cells are positive for SATB2 immunohistochemical stain (SATB2 x200).**

## DISCUSSION

Craniofacial osteosarcoma typically affects mature skeletal bone; nearly 80% of the cases are reported in patients older than age 20 years. The peak incidence of these osteosarcoma is during third and fourth decades of life with no gender predilection. When an osteosarcoma occurs in patients aged more than 40 years, it is usually secondary to Paget's disease of bone or irradiated bone lesions. There was no associated risk factor detected in this case.

An increased serum level of alkaline phosphatase (ALP),

which can occur in about half of the patients, reflecting an ongoing osteoblastic activity. The elevation of ALP level after surgery indicates persistent, recurrent or metastatic disease. In addition, those patients with sphenoid wing osteosarcoma commonly present with temporal bossing, temporomandibular joint pain, proptosis, facial mass, decrease in vision and headache. Conventional radiograph provides limited information regarding craniofacial osteosarcoma owing to the complex anatomical structure of the craniofacial bones. For assessment of lung and bone metastases, chest CT scan and radionuclide bone scan can be performed respectively (3). The bone windows CT scan is the most useful in aiding the diagnosis as lytic and periosteal remodelling are the common features. While the CT scan or MRI with contrast enhancement is effective for assessment of adjacent soft tissue involvement.

Based on the clinical history, radiological results, and site of the tumour, the most likely diagnosis includes meningioma, metastatic tumour or aggressive bone lesion such as osteosarcoma. However, histopathological result revealed the diagnosis of typical histomorphology of osteosarcoma. Since there is no evidence of other primary site of bone or soft tissue lesion elsewhere, no risk factors such as previous radiotherapy, fibrous dysplasia or Paget disease in this patient, we strongly believe that the most likely diagnosis for our case is primary osteosarcoma of sphenoid bone.

The majority of osteosarcoma cases present with features that are strongly suggestive of the diagnosis on the basis of patient's age, tumour location and typical radiographic appearance of aggressive bone destruction, tumour matrix mineralisation (either osteoid or chondroid), soft tissue involvement and periosteal bone formation. Though the current case shows classical morphology of an osteosarcoma, the diagnostic dilemma arises when the osteosarcoma presented in an older age group and involving a rare site.

Craniofacial osteosarcoma is microscopically similar to the long bones' osteosarcoma counterpart. The neoplastic cells display severe anaplasia and pleomorphism; epithelioid, plasmacytoid, fusiform, small and round or spindle shapes. A minimal osteoid formation by the malignant cells is necessary to render a diagnosis of osteosarcoma. The neoplastic bone which is varies in quantity and woven in architecture (rarely lamellar) is intimately associated with the tumour cells. It is deposited as disorganised trabeculae that may produce coarse lace-like pattern or broad, large sheets formed by coalescing trabeculae. The neoplastic bone is eosinophilic when unmineralised and basophilic if mineralised. The conventional osteosarcoma is subclassified according to the most-prominent cell type; osteoblastic, chondroblastic, giant cell rich, osteoblastoma-like, epithelioid, clear cell and chondroblastoma-like. SATB2 is a marker for osteoblastic differentiation. It is sensitive marker with sensitivity of 94% but is not specific with specificity of 55% (4). The immunoexpression of SATB2 is demonstrated in other high grade primary bone sarcomas.

Conventional osteosarcoma is highly an aggressive and lethal osseous neoplasm. Eradication of the primary tumour and elimination of the distant metastases are the goal of the therapy. Total surgical resection with tumour-free margin is the mainstay of the treatment. The positive resection margins are predictors for local recurrence and are associated with poor prognosis. However, a complete resection of the tumour is often not achievable in skull base osteosarcomas owing to the associated risk of surgical morbidity therefore often requiring adjuvant chemotherapy in most cases. Moreover, there is no consensus on safety margins when performing surgery on the skull. There was a study reported an improved in survival outcome with the use of chemotherapy. Radiotherapy is reserved for the unresectable tumours or tumour with incomplete resection. The general treatment for craniofacial osteosarcoma includes surgery, radiotherapy and chemotherapy. Having said that, the prognosis is not promising. In this case patient was not keen for chemotherapy and opted for radiotherapy and palliative care. The possibility of unchanged of the tumour size in this patient is might be due to unresponsiveness to radiotherapy but since the patient opted for the palliative care no further aggressive treatment was done.

The recurrence rate of skull osteosarcoma is high, 75%. However, distant metastases, typically to the lung and brain occur less frequently in head and neck osteosarcoma as compared to the other anatomical location in about 7% to 17% of the patients (5) .

## CONCLUSION

In conclusion, primary osteosarcoma of the skull base is a rare entity and extensive experience with this neoplasm is lacking. A correlation with the clinical and radiological findings is crucial in making the accurate diagnosis.

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