Infratemporal Fossa Osteosarcoma; A Rare Presentation
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Introduction
Osteosarcomas are the most common primary bone malignancies, accounting for 40% to 60% of all primary malignant neoplasms of bone. While osteosarcomas typically occur in the long bones, osteosarcomas of the head and neck are relatively rare, comprising about 10% of cases, usually occurring in the mandible or maxilla. These malignancies usually present in long bones during the second decade of life, compared to its presentation in the head and neck which typically occurs during the third to fourth decades of life. Patients who have undergone previous chemotherapy with alkylating agents or radiation therapy for another tumor have been found to have a higher incidence of occurrence of head and neck osteosarcomas. Paget’s disease, retinoblastoma, and benign bone diseases have also been found to be predisposing factors for this malignancy.

Osteosarcoma is characterized histologically by malignant spindle cells producing immature bone or osteoid. It is subdivided into chondroblastic, osteoblastic, and fibroblastic types, depending on the major histologic pattern present. The chondroblastic type has been the most frequently described for head and neck osteosarcomas.

In this article, we describe our experience with the extremely rare presentation of a chondroblastic osteosarcoma located in the infratemporal fossa of a pediatric patient at a tertiary care center and teaching hospital.

Case Report
A 17-year-old male presented with an infratemporal fossa mass causing nasal obstruction and Pressing his hard palate medio-inferiorly after having undergone total body radiation and bone marrow transplant for acute lymphoblastic leukemia three years prior. The mass was biopsied; histologic morphology and immunochemistry revealed chondroblastic osteosarcoma.

The patient underwent pre-operative radiation and chemotherapy in attempts to decrease the size of the mass but the tumor continued to increase in size despite treatments. The patient then underwent a salvage resection of the tumor. A bicoronal incision was carried down to the level of the zygoma. Osteotomies were then performed with removal of the zygoma. A cervical incision was used to access and then reflect a segment of the mandible. The tumor, originating in the infratemporal fossa, was then resected from the skull base. The maxillary division of the trigeminal nerve was incorporated in the mass and was sacrificed along with the tumor. The zygoma and the mandibular segments were then replaced to their normal anatomical positions and the surgical wounds were closed. The patient did well post-operatively.

The patient subsequently required cosmetic Gore-Tex implantation for temporalis muscle deficiency and obturator management of an oral-antral fistula. The patient then required two rounds of chemotherapy. The patient then underwent five rounds of chemotherapy. The patient then successfully underwent two rounds of chemotherapy. At two years post surgery the patient was disease-free.

Discussion
While osteosarcoma is the most common bone malignancy, the prevalence of its presence in the head and neck remains relatively low. Previous radiation or chemotherapy have been linked to increasing the risk of pediatric patient developing a secondary bone sarcoma. The primary location of malignancy has been reported to be the mandible, followed by the maxilla. The infratemporal fossa has not been previously reported as a location of this malignancy.

The treatment of choice for osteosarcoma remains surgical resection with wide margins. The efficacies of adjuvant therapies, such as chemotherapy, for head and neck osteosarcomas have been used with limited results. Osteosarcomas of the head and neck have also been found to have a higher incidence of occurrence of head and neck osteosarcomas.

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Pathology
Fig 1. H&E; 40x. Malignant spindled cells set within lobules of malignant cartilage.

Fig 2. Post-operative axial view showing resultant defect.

References