



# Primary Intramedullary Osteosarcoma of the Maxilla in a Pediatric Patient: A Case Report and Review of the Literature

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## Abstract

Osteosarcomas are the most common malignant neoplasms of bone; mainly found in long bones. However, osteosarcomas can occur anywhere in the body. Osteosarcomas rarely occur in the head and neck and are even more uncommon in the maxilla. Primary osteosarcomas of the maxilla in pediatric patients is extremely uncommon, as head and neck osteosarcomas usually present in the third to fourth decades of life. Head and neck osteosarcomas usually arise secondary to radiation or chemotherapy treatment of a preexisting tumor.

In this case report, a lesion was discovered upon routine radiographic and clinical exam in a pediatric dental office. Once the lesion was biopsied and the well differentiated, intramedullary osteosarcoma was confirmed histopathologically, a right hemi-maxillectomy was performed. Virtual surgical planning was based off a medical grade maxillofacial Computed Tomography (CT) scan. In operable cases, surgical resection of the tumor and surrounding tissue is the gold standard of treatment. Virtual planning allows for precise and accurate surgical margins. After the patient's case was reviewed by the Joe DiMaggio children hospital tumor board, a consensus agreement on treatment planning deferred post-surgical systemic chemotherapy due to possible long-term complications.

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## Introduction

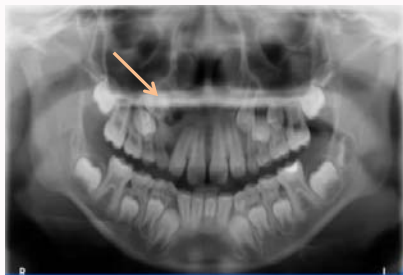
Osteosarcoma is a primary malignant neoplasm arising from mesenchymal stem cells, most commonly found in the long bones around the knee [1,2]. Maxillofacial osteosarcomas are rare and constitute a minor percentage of all diagnosed head and neck tumors [3]. Studies show that the maxilla is less commonly affected than the mandible. Upon occurrence, maxillary osteosarcomas arise more commonly in the alveolar ridge and the maxillary antrum. In contradistinction to osteosarcomas of long bones, which demonstrate a bimodal age distribution, the peak incidence for jaw osteosarcoma is most commonly observed during the 3<sup>rd</sup> to 4<sup>th</sup> decades [2,4]. This case study examines the incidence of an osteosarcoma of the maxilla diagnosed in an 8-year-old male and analyzes treatment modalities.

## Case Presentation

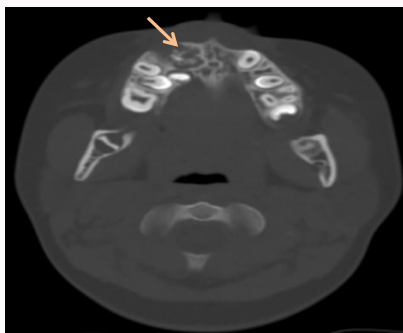
An 8-year-old African American male presented with his mother to Nova Southeastern University Department of Oral and Maxillofacial Surgery with a history of diffuse swelling above his upper right front teeth consistently for two months. The patient reported that he was asymptomatic with no pain, no numbness, nor any history of fever, loss of appetite, weight loss, epistaxis, nasal obstruction, or change in vision. There was no contributory medical, social or family history. Extra-oral examination was unremarkable; no palpable lymph nodes at any level of the head and neck region were clinically detected. Intra-oral examination revealed a firm 3 cm × 3 cm solitary swelling overlaying the buccal aspect of the right maxillary alveolar ridge at the deciduous tooth B, extending anteriorly near to the incisors, but not crossing the midline (Figure 1). There was expansion of the buccal cortices, however no palatal expansion was observed. Orthopantomograph (OPG) revealed a poorly defined mixed-density lesion of the upper right maxilla denoting irregular areas of osteolysis with Garrington's sign in relation to the upper right impacted canine (Figure 2). Furthermore, a CT scan with intravenous contrast was taken, and axial, coronal and sagittal cuts revealed poorly defined areas of sclerosis with few lytic areas seen involving the right maxillary sinus causing



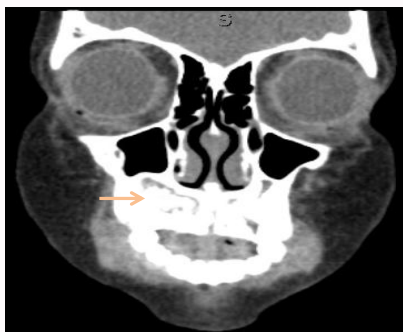
**Figure 1:** Intraoral Image showing clinical appearance of solitary lobulated swelling at the right maxillary alveolar ridge (arrow).



**Figure 2:** Orthopantomograph (OPG) showing an ill-defined mixed radiolucent-radiopaque lesion in the right upper jaw (arrow).



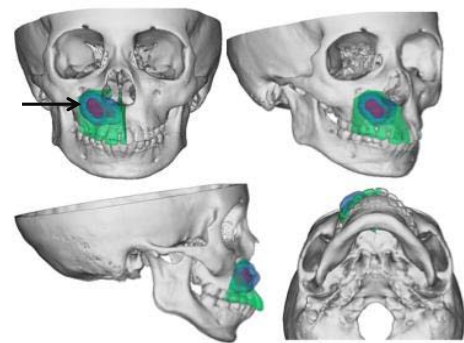
**Figure 3A:** Axial view of computed tomography demonstrating an expansive, ill- defined mass centered to the right maxillary alveolar process, destructing the bone (arrow).



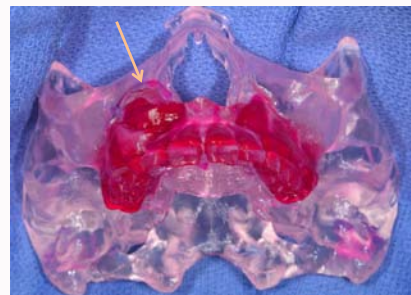
**Figure 3B:** Coronal view of computed tomography demonstrating an expansive, ill- defined mass centered to the right maxillary alveolar process, destructing the bone (arrow).



**Figure 3C:** Sagittal view of computed tomography demonstrating an expansive, ill- defined mass centered to the right maxillary alveolar process, destructing the bone (arrow).



**Figure 4A:** A right hemi-maxillectomy was planned using Virtual Surgical Planning (VSP) and the lesion was identified (arrow).



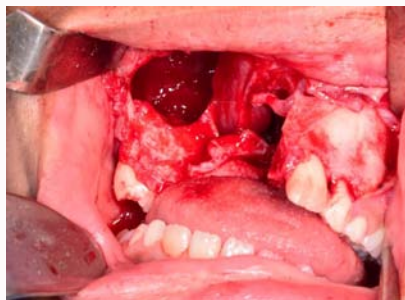
**Figure 4B:** The stereolithographic surgical model was fabricated and the lesion was identified (arrow).

disruption of normal architecture of the involved bone (Figure 3A-3C). Initially, an incisional biopsy of the right maxillary lesion was performed under general anesthesia. Aspiration was negative for

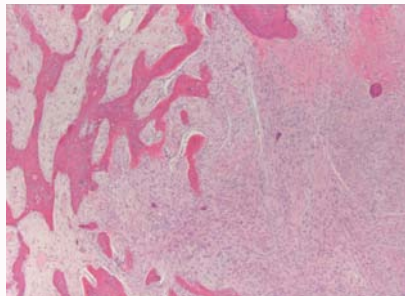
fluid. Perioperatively, the lesion appeared as fibrous tissue with granulation. The initial biopsy revealed osteoid tissue with irregular trabeculae, with osteoblasts appearing large round to oval, epithelioid in appearance with hyperchromatic nucleus and prominent nucleoli and occasional atypical mitoses. Intertrabecular areas showed solid cellular stroma. The tumor exhibited an aggressive infiltrative pattern with diffuse margins. The biopsy was sent to multiple pathologists, who collectively agreed that the lesion represented one of the three differential diagnoses: 1) Desmoplastic fibroma; 2) Well-differentiated intramedullary osteosarcoma; or 3) Atypical fibroma, aggressive in nature. A right hemi-maxillectomy using virtual surgical planning was performed. The lesion was identified using cutting guides and stereolithographic surgical models were fabricated (Figure 4A and 4B). The patient was taken to the operating room for resection of the tumor with at least 1 cm clearance around all the margins (Figure 5A and 5B). The right maxilla was removed in entirety up to the mesial



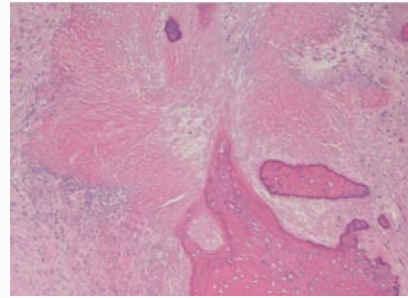
**Figure 5A:** Final surgical specimen showing the resection of tumor with bony margins of 1 cm.



**Figure 5B:** Right hemi-maxillectomy for resection of the tumor with at least 1 cm clearance at all the margins.



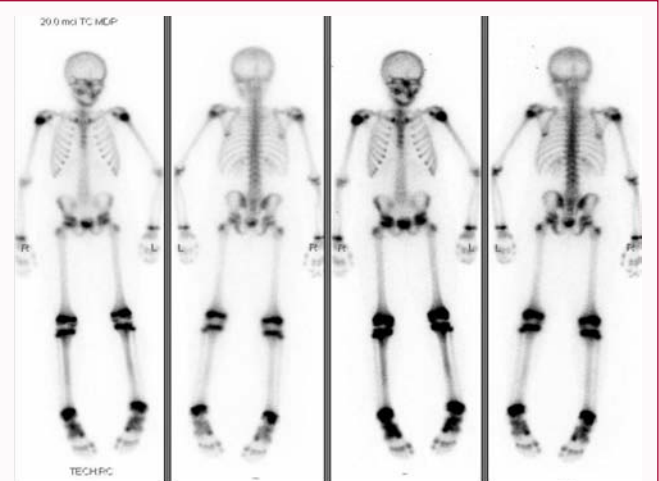
**Figure 6A:** Histology of well-differentiated osteosarcoma. Photomicrographs show a degree of cellularity that varies from acellular zones to hypercellular spindle cells fascicles. Most of the tissue is composed of hypocellular bundles of very dense collagenous tissue, with small spindle-shaped nuclei, reminiscent of desmoplastic tumor of bone. (Hematoxylin and Eosin stain, original magnification  $\times 400$ ).



**Figure 6B:** Mitotic activity and giant cells are absent, but vital bone spicules are present at the borders of the specimen. (Hematoxylin and Eosin stain, original magnification  $\times 400$ ).



**Figure 7A:** A 12-month postoperative Magnetic Resonance Imaging (MRI) of this case shows no mass or abnormal recurrent lesions at anterior maxilla.



**Figure 7B:** A 12-month postoperative total body bone scan of this case shows no evidence of recurrence.

of the first molar. Histopathologic report confirmed the diagnosis of spindle cell neoplasm best classified as well-differentiated low grade intramedullary osteosarcoma (Figure 6A and 6B). The patient was referred to follow up with a pediatric oncologist who recommended observing patient at 3-month intervals.

## Discussion

Osteosarcomas are malignant bone tumors characterized by formation of disorganized immature woven bone or osteoid tissue from mesenchymal tumor cells [1]. The management of all variations of osteosarcoma, including low-grade, intramedullary, and juxtacortical osteosarcomas, are identical with complete surgical excision of the tumor, in conjunction with wide surgical margins [5]. In cases of osteosarcoma of the jaws, sufficient surgical treatment at

an early stage is a key determinant of favorable outcome. Okinaka and Takahashi reported a case in which incomplete surgical resection of maxillary osteosarcoma ultimately led to distant metastasis to the lungs [6]. The report found that distant metastasis of maxillary osteosarcoma almost exclusively involved the lungs, and that distant metastasis presented at a later time than expected given the aggressive nature of the tumor. Osteosarcomas arising from the maxilla cannot always be resected with sufficiently broad margins to ameliorate patient safety [6]. Therefore, surgical therapy is often supplemented with adjuvant treatment to prevent recurrence. Kupeli et al. [3]



reported a case of pediatric maxillary low-grade osteosarcoma which was subsequently treated with surgical resection, followed by chemotherapy. Histopathologic assessment of the resected tumor revealed positive margins. Due to high risk of recurrence, six courses of chemotherapy consisting of Cisplatin (120 mg/m<sup>2</sup>) and Doxorubicin (30 mg/m<sup>2</sup>) at 3-week intervals was prescribed. After chemotherapy treatment was completed he patient showed no signs of residual or recurrent tumors, and was subsequently in remission thereafter [3]. Although the use of chemotherapy is controversial it can be advantageous in its use as adjuvant treatment in cases where there is a high risk of recurrence; specifically, tumors >4 cm, positive surgical margins, young age, and previous diagnosis of retinoblastoma [3]. Consequently, due to the lack of high risk factors in this case, it was decided that adjuvant chemotherapy was not necessary. The patient would be closely monitored for any suspicious changes [7].

Furthermore, cases of post-operative radiotherapy have been cited as possible adjuvant treatment for osteosarcoma of the jaws. Prabhusankar et al. [8] reported a case of maxillary osteosarcoma, which approximated the orbital region in a 20-year-old male. The resected tumor showed positive margins along the superior aspect. To protect the orbital region during resection, post-operative radiotherapy was performed [8]. Radiotherapy as adjuvant treatment is particularly advantageous if surgical resection is incomplete, as seen in the aforementioned case [8]. Review of the literature revealed that incidence of osteosarcoma of the maxilla in the first decade is a rare entity and usually behaves differently from the osteosarcoma of other bones. These lesions appear to grow slowly and may be observed after surgical resection [9-12]. A 12-month postoperative imaging of this case revealed no recurrent lesions (Figure 7A and B). Patient regularly follows up with a pediatric oncologist every three months. In summary, children and adolescents of either gender are susceptible to developing primary non-radiation-associated osteosarcomas in the head and neck region, most commonly arising in the mandible. These patients generally present with symptoms of swelling and a mass of short duration, although dental problems and paresthesia have also been observed. After radiographic examination, complete surgical extirpation followed by appropriate adjuvant therapy for osteosarcoma will allow pediatric patients to demonstrate a better overall prognosis, irrespective of gender, anatomic location, size of tumor, or tumor histology. According to Demicco et al. [7] well-differentiated gnathic osteosarcoma exhibits a relatively low-grade malignancy and has an excellent prognosis with wide local excision. If possible, radiation therapy should be avoided to prevent long-term growth stunting complications due to the patient's susceptibility to growth disturbances in pre-pubescent age [7].

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