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Childhood Osteosarcoma of Mandible: A Rare Case Entity

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Running title: Childhood Osteosarcoma

Abstract: ***Osteosarcoma (OS) of the jaws is a relatively rare malignant bone tumor. Like, its counterpart in the long bones, OS affecting the head and neck region shows distinct yet diverse clinical, histologic and prognostic characteristics. Here, we report a rare case of OS in a 12-year-old child, who came with a bony swelling in the right mandibular posterior region, with a radiographic presentation of sunburst appearance, the histopathological examination confirmed the diagnosis. The patient underwent wide local excision with hemimandibulectomy followed by free fibula flap reconstruction under general anesthesia and is currently undergoing regular follow-up examination.***

Keywords: osteosarcoma, head and neck, free fibula flap

1. Introduction

Majority of all oral cancers are squamous cell carcinoma, which are epithelial tumors. However, various primary tumors can be seen in head and neck region, and they are either epithelial, mesenchymal or hematolymphoid[1]. The term osteosarcoma refers to a heterogenous group of primary malignant neoplasms affecting bone forming or mesenchymal tissues that have histopathologic evidence of osteogenic differentiation[2]. About 10% of osteosarcoma occurs in the head and neck region, with jaws affected most frequently with slight predilection for the mandible[3,4].

Early diagnosis and surgical resection with wide margins are important to obtain local control of the disease and to improve the prognoses of patients with head and neck osteosarcomas[5]. Therefore, surgical resection is often followed by adjuvant radiation therapy and/or chemotherapy to prevent recurrences[6].

We present a rare case of osteosarcoma of the mandible of a child who was treated surgically with micro-vascular reconstruction with free fibula flap followed by neoadjuvant chemotherapy The uniqueness of this case lies in the rarity of the patient's age and site of this disease leading to a difficulty in diagnosis along with its management for achieving better functional and cosmetic results.

2. Case Report

A 12-Year-old healthy male child from Tajikistan referred to the Department of Surgical Oncology, Dharamshila Narayana Super-speciality Hospital, New Delhi with a history of swelling in right side of lower jaw since 6 months. His parents were concerned and stated that the swelling began growing rapidly involving the cheek. During this time, patient had undergone incisional biopsy at Tajikistan which revealed Conventional Osteosarcoma. The patient had no history of trauma or any other disease affecting the jaws or any other systemic illness.

Upon his visit, clinical examination and further investigations were done. On examination, a hard, well-defined, non- tender swelling was detected on Right side of the body of the mandible with bucco-lingual expansion lesion of right lower mandible extending from canine to Retro-molar pad area following loosening of teeth. Tumour extends to the overlying soft tissues with no evidence of paraesthesia and lymph node involvement.

A Contrast Enhanced MRI scan of the face and neck showed an expansile enhancing lesion of size $3.62 \times 4.37 \times 4.13$ cm in the region of alveolar process and body of right side of the mandible with significant soft tissue component and destruction of the outer cortex (buccal component) suggestive of any of the following: Chondromyxoid fibroma, Desmoplastic ameloblastoma, Ameloblastic fibroma/ carcinoma and Osteosarcoma.

Slide and Block review of the biopsy specimen was performed. Microscopic examination demonstrated pleomorphic polygonal and spindle cells with mitoses and foci of lacy lesional osteoid with abundant cartilaginous matrix permeating host bone suggestive of Grade- III (High grade) Chondroblastic type of Osteosarcoma.

The case was discussed with multimodality tumour board and in view of the patient's age and aggressive behaviour of the disease, it was decided to treat him with a curative intent. The treatment plan includes Wide Local Excision of the tumour with Right side Hemimandibulectomy followed by Free Fibular Flap reconstruction. Under General anaesthesia, transverse neck incision was made and surgical resection with hemimandibulectomy done preserving the condyle for its functional stability. The surgical sample obtained sent for frozen sections showed macroscopic and microscopic surgical margins free of tumour. The case was then handed over to Plastic Surgery team where Right Fibular Flap of 12×3 cm was harvested and osteotomy cuts were made 3cm to make ramus and a 6 cm to make body of the mandible and fixed with 2 mm plates and screws. Microvascular anastomoses were performed on Right Neck with Right Facial Artery and branch of Right Internal Jugular Vein and directly with External Jugular Vein. Flap interpolated and intra- oral flap insetting done with anatomical closure of the neck wound. Post-OP recovery was uneventful.

Based on the post-operative histopathology report, adjuvant chemotherapy was recommended and he received one cycle of OGS-2012 based chemotherapy at 2-weeks of postoperative period. During subsequent follow-ups, the surgical site was completely healed and no other local abnormalities has been ruled out.

3. Discussion

Literature suggests OS of jaw differs from the OS of the long bones in its biological behaviour, presenting a lower incidence of metastasis and a better prognosis with approximately 40% 5-year survival rate as compared to 20% for non-jaw lesions[7,8]. Usually, OS of jaw present in third and fourth decades of life almost a decade after their presentation in long bone tumors. However, in the present case we dint find any such as the patient was very young[9].

The etiology of primary type is unknown; may be due to genetic influence or other environmental factors[10]. Various predisposing factors of osteosarcoma include hereditary retinoblastoma, Paget's disease of bone, a history of fibrous dysplasia, or trauma[3, 11].

Generally, the clinical picture of osteosarcomas in head and neck region are discreet and include regional swelling, low-intensity pain, paraesthesia, changes in tooth position, loose teeth, and changes in the fit of prosthesis. Involvement of temporomandibular joint or para mandibular musculature is often accompanied by trismus[3,4].

The radiological characteristics ranges from osteolytic to mixed osteogenic (appearance of sun beams) pattern of bone[12]. Additionally, there is an association between oral osteosarcomas and some radiographic images with a "sun-ray pattern"[9]. Magnetic Resonance Imaging can play a central role in the interpretation of extent of bone marrow invasion showing soft tissue mass with enlargement of the periodontal ligament space and destruction of cortical boundaries. This finding was also seen in our case[10].

Consequently, knowing the radiological and clinical properties of malignant lesions will provide the determination of biopsy necessity and type followed by histopathological analysis for definitive diagnosis[13]. Among the histological subtypes, the chondroblastic variant is considered to be highly prevalent in head and neck region and associated with a better prognosis[4,14]. Chondroblastic type, consists of atypical chondroid areas composed of pleomorphic binucleate cells and fibroblastic type, shows atypical spindle shaped hyperchromatic cells[8]. The present case exhibits feature of High- grade chondroblastic variant of OS. Previous studies have found immunohistochemical expression of p53, MDM2, CDK4, PCNA and ki67 proteins as favourable prognostic markers[15]. In this case, immunoreactive score for SATB-2- [EP281] ad Vimentin was positive in neoplastic cells.

The treatment of choice for osteosarcoma is complete surgical resection with wide margins, although chances local recurrence is more common due to not only in difficulty of obtaining clear margins but because of complex anatomical limitations in head and neck region. This has resulted in several interdisciplinary treatments based with surgical resection followed by adjuvant radiotherapy/ chemotherapy to prevent recurrences[6]. The prognosis of patients with OS has been reported to be statistically better with these factors: absence of paraesthesia as a symptom, smaller tumor size, adequacy of surgical removal, a more differential histological grade of the lesion and younger age of the patient, increase age may be related to recurrence of the treatment[9].

Osteomyelitis of jaws in children is a rare entity and prompt diagnosis with rapid management by combination of surgery and chemotherapy is the ideal treatment of choice with good functional and cosmetic outcome.

Contributor name	Contribution
Dr. Anshuman Kumar	Concept, manuscript review
Dr. Ehsan Siddiqui	Manuscript review, literature search
Dr. Nilotpal Mishra	Manuscript preparation, manuscript editing, literature search
Dr. Birsubhra Roy-guarantor	Manuscript preparation, manuscript editing, literature search

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Figure Legends

Fig 1-Pre-operative intra-oral picture of the lesion
Fig 2-Pre-operative extra-oral swelling
Fig 3- Computed tomography
Fig 4-Resected Specimen
Fig 5-Intra-operative picture after resection
Fig 6-Harvested Free fibula flap with Recon plate
Fig 7-Post-operative extra-oral image