



Clinico-Radiographical Findings in Mesenchymal Chondrosarcoma of Maxilla – A Rare Case Report

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Authors' contributions

This work was carried out in collaboration among all the authors. Concept and designing of the manuscript was done by authors AS. Editing and academic analysis was collectively done by authors RG and NS. Literature review and grammatic correction was done by author SSS. All Authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Chondrosarcoma is classified as a malignant tumor formed by the abnormal differentiation of hyaline cartilage. A painless, slow growing swelling with a lobulated appearance is the most common clinical feature of Chondrosarcoma. Mesenchymal Chondrosarcoma is a rare high grade histological variant of chondrosarcoma. There is skeletal and extra skeletal manifestation of this tumor and has a relatively poor prognosis. In this article we are discussing a case report on mesenchymal chondrosarcoma involving maxilla and the pterygopalatine space. A brief review on radiological features and differential diagnosis is also included in this article.

Keywords: Mesenchymal chondrosarcoma; maxillo-facial radiology; pterygopalatine space.

1. INTRODUCTION

Mesenchymal chondrosarcoma is a subset of chondrosarcoma, which arises from the

remnants of the embryonic cartilage or metaplasia of meningeal fibroblasts. The mesenchymal form is a rare high grade variant of chondrosarcoma, which constitutes only about 2-

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3% of chondrosarcoma [1]. It has a slight female predilection and was first reported by Lichtenstein and Bernstein in 1954. It is seen most commonly in children and young adults within the age range of 15 to 35 years. The patient usually presents with long duration of swelling with a vague history of pain. In jaws it is more commonly seen in the posterior maxillary region. The tumor has both skeletal and soft tissue manifestation (33%) and has a relatively poor prognosis [2].

2. CASE REPORT

A 19-year-old female patient reported to our OPD with a history of painless swelling in the right cheek region since one year [Fig. 1]. The swelling started developing after being hit by a ball while playing and was progressively enlarging since then. There was no history of any deleterious habits and no similar swelling was found on any other part of the body. The medical and family history was non-contributory.



Fig. 1.

On extra oral examination, there was a diffuse ovoid smooth swelling on the right maxillary region of size approximately 6x5 cm extending superiorly from the infraorbital region, inferiorly to

the ramus of mandible region, medially from the right nasolabial fold extending laterally to the zygoma. There was obliteration of the nasolabial fold, lagophthalmos and drooping of commissure of the left lip, while diplopia was absent. On palpation the swelling was bony hard with no crepitus present. Sensory deficit was not elicited. The skin over the swelling was normal and there were no palpable regional lymph nodes [Fig. 2].



Fig. 2. Ovoid swelling in maxilla extending to infraorbital and posterior zygoma region

Intra Oral Examination showed expansion of the palatal and the buccal cortical plate in relation to 13 to 18. There was no tenderness on palpation. The mucosa overlying the swelling was normal and there was no local rise in temperature. There was no tenderness on percussion or mobility with respect to the associated teeth. There were no carious teeth or periodontal pathology present at the same side of the jaw [Fig. 3].

Based on the clinical evaluation, we made a provisional diagnosis of Primary or Metastatic malignant tumor of the jaw with differential diagnosis of Fibrous dysplasia and Odontogenic cysts or tumors of the jaw.

On Radiological examination IOPAR 14,15,16,17 region showed ill-defined rarefaction and altered

trabecular pattern in relation to root apex of 15, 16,17 also causing resorption of the roots of 15 in the apical third and 16,17 upto the middle third. Maxillary occlusal topographic view showed mixed radiolucent-radiopaque mass with the expansion of the buccal alveolar plate in the posterior aspect of right maxilla [Figs. 4 and 5]. The panoramic radiograph showed a mixed radiolucent- radiopaque pattern involving almost the entire maxilla of size 6 x6 cm with faint cortical borders, there was also a soft tissue shadow present internally, also overlapping the right coronoid process of mandible. The tooth 18 was displaced in the maxillary sinus region. The lesion also seemed to extend into the inferior and lateral border of orbit [Fig. 6].

CECT revealed ill defined expansile lesion in the right maxillary region of size, measuring 6.5 x6.8 cm, completely occluding the right maxillary sinus, right nasal cavity and displaced the nasal septum towards the left [Fig. 7a]. It was also invading the ethmoid sinus. Superiorly, the mass has involved the floor of the orbit with infiltration to the right inferior rectus and inferior oblique muscle. Anteriorly it was infiltrating the buccinator space and right masseter [Fig. 7f]. Laterally the mass was involving the right pterygopalatine fossa causing rarefaction in the lateral pterygoid plate, but the medial pterygoid plate was well maintained [Fig. 7d]. The

expansile lesion was also having coarse calcification and soft tissue component in it. However, there was no intracranial extension seen. The radiological differential diagnosis was given as Chondrosarcoma, Osteosarcoma, and Squamous Cell Carcinoma.



Fig. 3. Expansion of the cortical plates

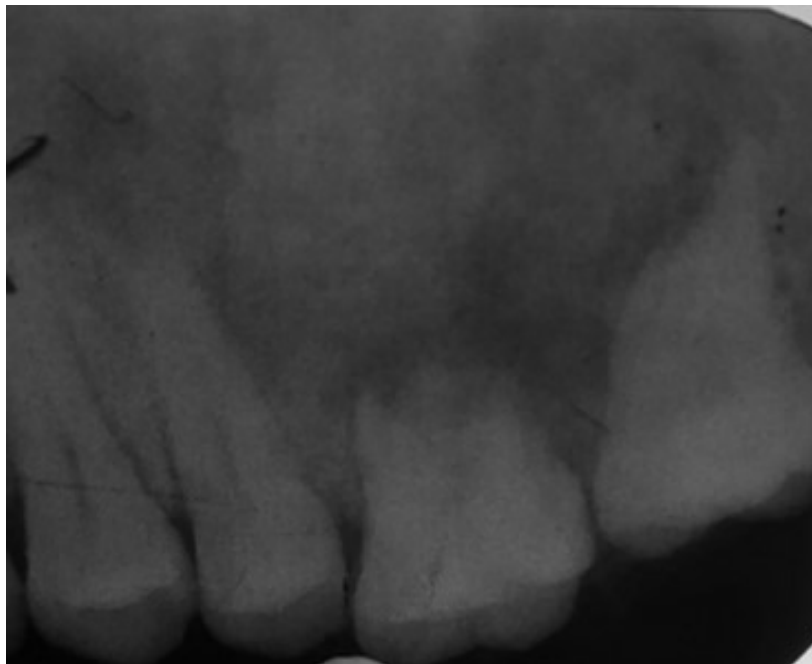


Fig. 4. IOPAR 15,16,17 region

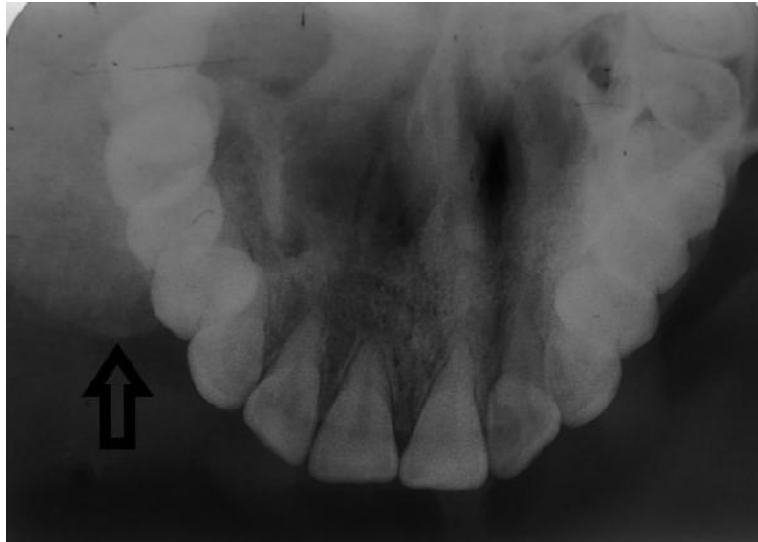


Fig. 5. Maxillary occlusal view

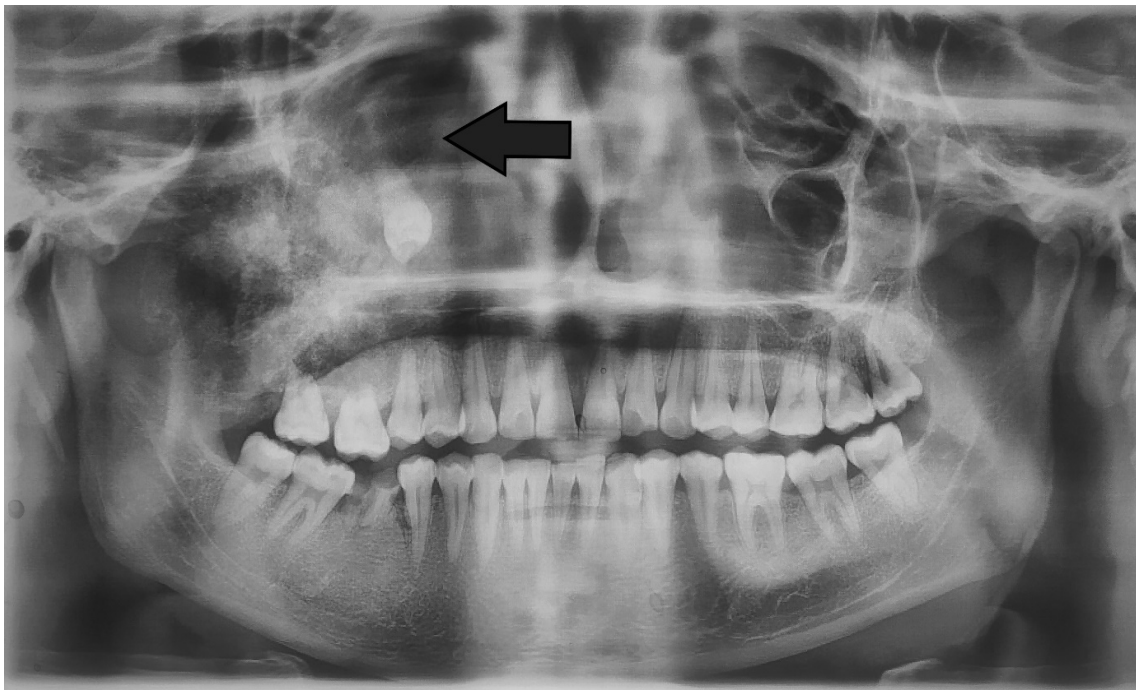


Fig. 6. Panoramic radiograph

Routine blood investigation showed normal blood Counts. Fasting blood glucose level was 96mg/dl. AST, ALT and Alkaline phosphatase were (27 U/L, 21U/L & 51U/L respectively) within the normal biological reference range. Incisional biopsy was done and tissue measuring 2.6 x1. 3 cm was obtained. Histopathological examination

revealed small round or ovoid cells with hyperchromatic nucleus, inconspicuous nucleoli and scant amount of cytoplasm. There were lobules of chondroid tissue with mild pleomorphism. The histopathological report was suggestive of mesenchymal chondrosarcoma [Fig. 8].

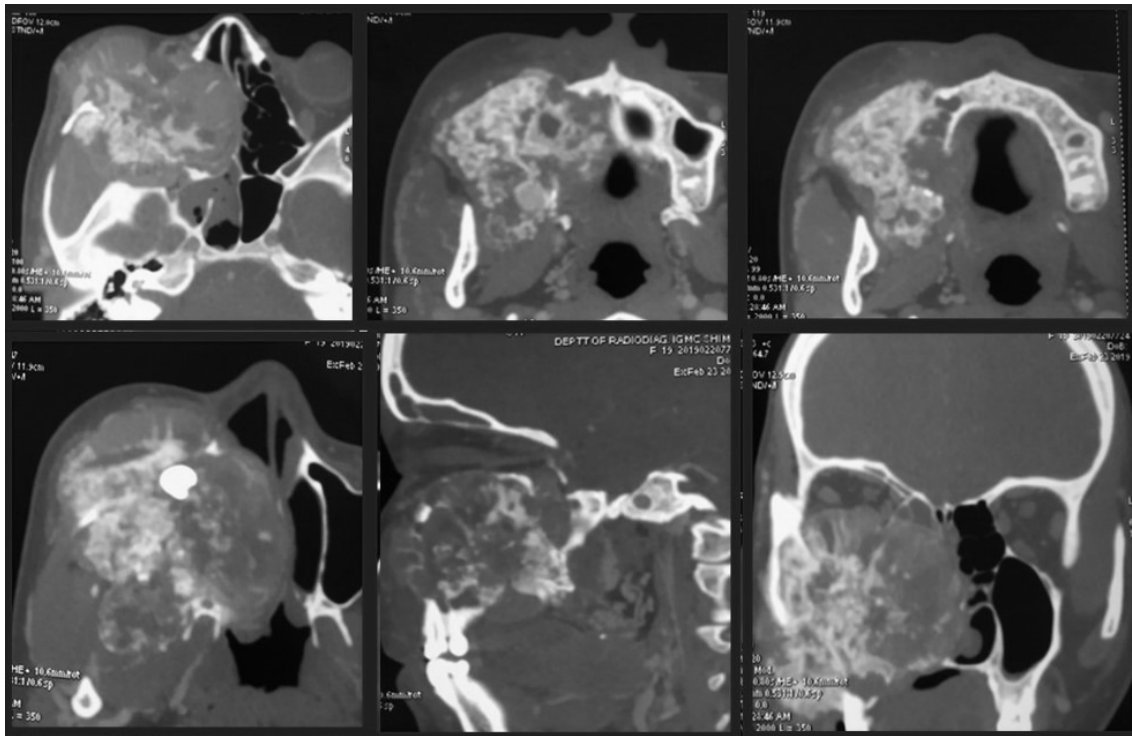


Fig. 7. CECT PNS: Axial (a,b,c,d), Sagittal (e) and Coronal View (f)

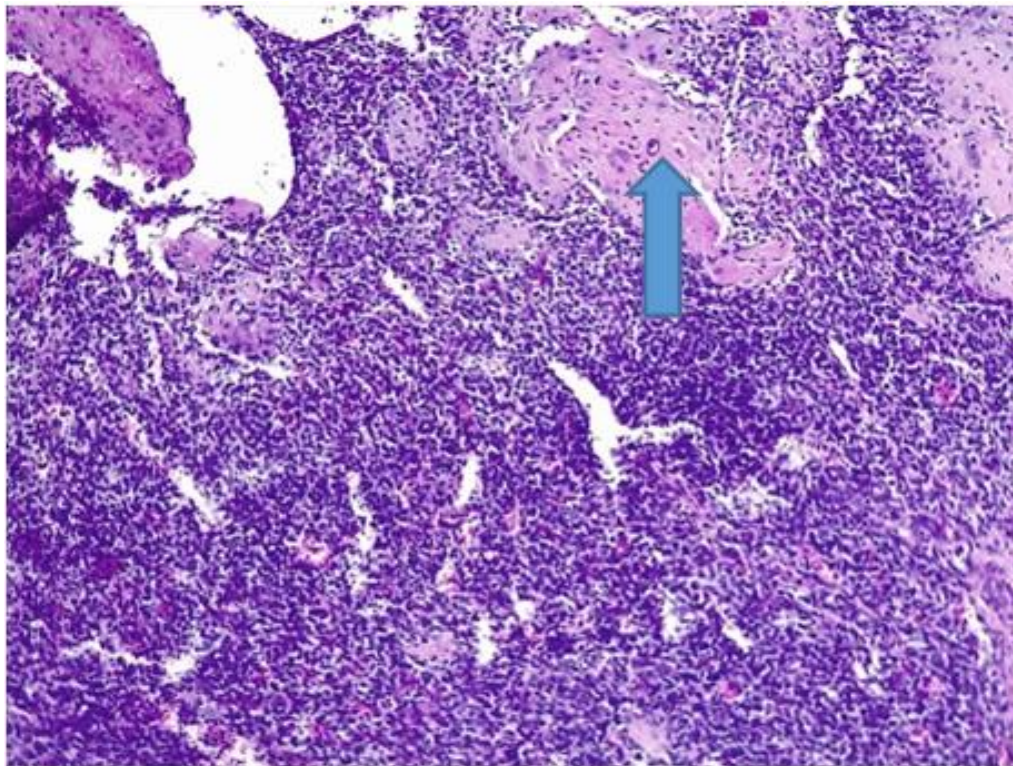


Fig. 8. Histopathological section showing chondroid tissue



Fig. 9. Post operative follow up with obturator

Based on the above findings a final diagnosis of mesenchymal chondrosarcoma was given. The patient was referred to the Department of ENT and the lesion was surgically excised, followed by chemo and radiotherapy. A removable partial denture with obturator was given post surgery [Fig. 9]. The patient was recalled after an interval of one year. The radiographic evaluation of Jaw bones and Chest X-Ray revealed a grossly normal study [Fig. 10].

3. DISCUSSION

Chondrosarcoma is the third most common malignant tumor of jawbone [1]. According to WHO Chondrosarcoma is classified as a malignant tumor formed by the abnormal differentiation of hyaline cartilage. The etiology of chondrosarcoma is unclear and chondrosarcoma of the jaw is very rarely published in the literature. A painless, slow growing swelling with a lobulated appearance is the most common clinical feature of chondrosarcoma. It is more commonly seen in the maxilla than in the mandible where as an extension to posterior pterygopalatine space is seen very rarely [3]. Various forms of Chondrosarcoma are discussed in Table 1.

Mesenchymal chondrosarcoma is an aggressive and a high grade tumor with various skeletal and extra-skeletal manifestation including the Skull, Jaws, Orbit, CNS, Kidney, Spleen and Intracranial tumors [4]. Radiological features of

chondrosarcoma suggest two different patterns. A frank radiolucency in the early stage or a radiolucent lesion with various forms of radiopaque shadow which may be due to the deposition of calcified masses. It can also produce a band like widening of the periodontal ligament [1]. As per the literature various forms of cortical reactions like the Sun-ray appearance, Codman's triangle, Onion peel appearance are seen radiographically in osteosarcoma and chondrosarcoma [1]. A brief description of radiological differential diagnosis is given in Table 2. Histologically the tumor shows a biphasic pattern consisting of both small cells and islands of atypical cartilage [4]. The tumor is highly invasive with delayed metastasis to other bones, lymph nodes and the lungs. Xu J et al. [5] in 2015 published a systematic review of mesenchymal chondrosarcoma involving the bone and soft tissue published data of 107 patients in the past 20 years was analyzed and concluded that 5-, 10- and 20-year overall survival is 55.0%, 43.5% and 15.7% respectively. The tumour also shows high chances of recurrence. There is no evidence based treatment protocol for the chondrosarcoma of the jaw. However a multimodal approach is followed, which includes surgery, chemotherapy, radiotherapy, immunotherapy and combination of various treatment modalities. A surgically clear margin is essential for a favourable prognosis and to avoid recurrence. Adjuvant chemotherapy and radiotherapy has shown better prognosis and favourable outcome [6].

Table 1. A brief description of various forms of chondrosarcoma

Types	Clinical Features	Radiological Features	Prognosis
Periosteal chondrosarcomas [7]	1–2% of all cases of chondrosarcomas. Slow growing, well differentiated mass. The cortex can be eroded or thickened but never destroyed	Disseminated or localized ring like calcification. Tumoral nodules/satellitenodules seen	Favorable outcome after surgery
MesenchymalChondrosarcoma [8]	2–3% of all chondrosarcomas. Slow growing lobulated painless swelling. A biphasic pattern with areas of undifferentiated small round cells and well differentiated hyaline cartilage.	Expansile lytic lesion with coarse calcification. Extra osseous or soft tissue involvement often seen.	Prognosis is poor with early pulmonary and lymph node involvement. Ten year survival rate is 30-40%.
Clear cell chondrosarcoma [9]	(2%) of chondrosarcoma, low grade tumour , lytic epiphyseal with extension into metaphysis, Margins are well defined mostly. Tumour may remain unnoticed for year and may be noticed incidentally or after causing pathologic fracture.	Lobulated margin, calcified matrix not always present. There may not be an extension into soft tissue.	Good prognosis with five year survival rate in 92% cases.
Dedifferentiated chondrosarcoma [10]	10–12% of all chondrosarcomas, Pain, swelling, pathologic fracture is commonly seen.	Bimorphic pattern with lytic area and calcification. Huge soft tissue lesion with calcification is also indicative	Very poor. Overall five year survival rate is only. 8.5-13%
Secondary Chondrosarcoma			
A). Olliers Disease [11]	A rare bone disease that is characterized by multiple enchondromatosis with a typical asymmetrical distribution and confined to the appendicular skeleton	Asymmetrical osteolytic lesions with well-defined/ sclerotic margins is seen	Not well described
B) Maffuci's Syndrome [12]	A rare disease involving multiple enchondromatosis and cavernous hemangiomas of the dermis, subcutis, or internal organs. Enchondromas can lead to deformity or fractures.	Enlarging exostosis, the appearance of a less mineralized zone in the cartilage cap, calcifications in the soft tissues, the thickening of the cap (>1 cm) on CT and MRI suggest sarcomatous transformation	5-year survival rate of 25–35 %

Table 2. Gives a brief description of radiological differential diagnosis [1,6]

SL No.	Lesion	Predominat site	Additional features	Radiologic Appearance
1	Squamous Cell Carcinoma	Mandible> Maxilla	History of deleterious habit, Rapid growth metastasis, lymph node involvement	Radiolucency with radiopaque foci, bony sequestration seen
2	Metastatic tumor of jaw	Mandible> Maxilla	Unpredictive growth, Signs and symptoms of primary tumor	Solitary/ Multiple lytic lesion. Geographic/permeative pattern
3	Osteosarcoma	Mandible> Maxilla	Variable course and can cause necrotic slough in later stage	Radiolucency with radiopaque foci, sunburst appearance, widening of the PDL space
4	Chondrosarcoma	Maxilla> mandible	Lobulated appearance and slow growing	Onion peel growth of periosteal bone , codman triangle and cumulus cloud appearance
5	Odontogenic ghost cell Carcinoma	Cases rarely reported in literature	Variable	Destruction and opacification of jaw bone , ill defined area with absence of cortical reaction.

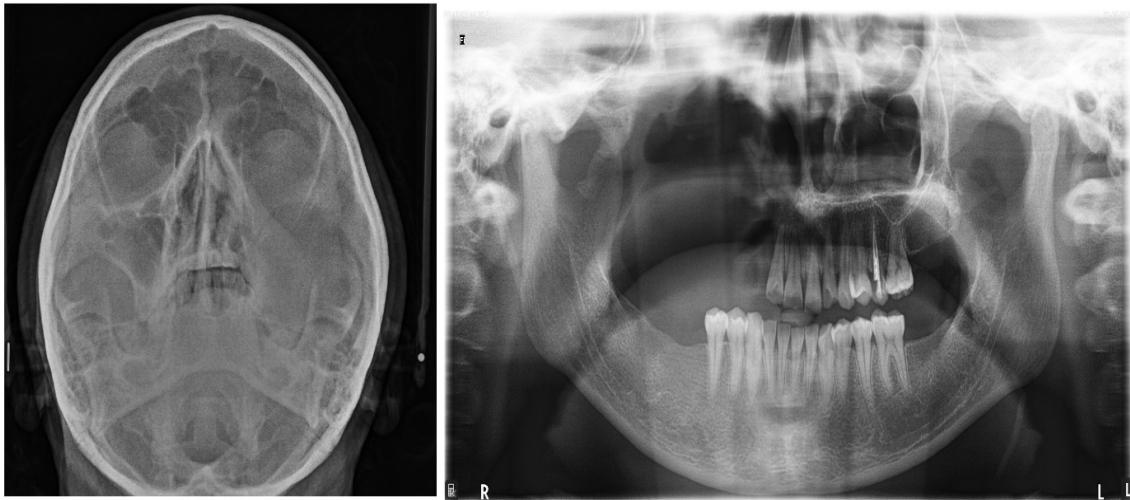


Fig. 10. One year follow up paranasal sinus view and panoramic radiograph

In this case, apart from the clinical aspect showing a slow growing tumour, coarse calcification with soft tissue component seen on the radiographs were strongly suggestive of sarcomatous changes. However the site of tumor involving the maxilla is more commonly seen in chondrosarcoma than osteosarcoma.

4. CONCLUSION

Mesenchymal chondrosarcoma is a high grade tumor, although very rarely reported in the literature, it can be included among the differential diagnosis of jaw tumors. The metastasis of lesion to another part of the body will further worsen the prognosis. The various radiographic features help in the early diagnosis and prompt treatment planning. The tumor in this case might have spread to the intracranial region if there was a delay in diagnosing and scheduling treatment.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Wood NK, Goaz W. Differential diagnosis of maxillofacial lesions Elsevier Publication 5th edition. 368-370.
2. Sammartino G, Marenzi G, Howard MC, Minimo C, Trosino O, Califano L, Claudio PP. Chondrosarcoma of Jaw: A close look at its Management. *Journal of Oral and Maxillofacial Surgery*. 2008;66(11):2349-2355.
3. Satomi T, Kaneko T, Abukawa H, Hasegawa O, Watanabe M, Matsubayashi J, Nagao T, Chiba H, Chikazu D. Chondrosarcoma of the maxilla extending to the pterygomandibular space: A case report and review of the literature. *J Maxillofac Oral Surg*. 2015;14(1):133-137.
4. Ollivier L, Vanel D, Leclère J. Imaging of chondrosarcomas. *Cancer Imaging*. 2004; 4(1):36-38.
5. Xu J, Li D, Xie L, Tang S, Guo W. Mesenchymal chondrosarcoma of bone and soft tissue: A systematic review of 107 patients in the past 20 years. *PLoS One*. 2015;10(4):e0122216.
6. Langlais RP, Langland OE. Diagnostic imaging of jaws Williams & Wilkins Publication 1st edition. 39-40.
7. Vanel D, De Paolis M, Monti C, Mercuri M, Picci P. Radiological features of 24 periosteal chondrosarcomas. *Skeletal Radiol*. 2001;30:208-12.
8. Justin Q. Mesenchymal Chondrosarcoma of the Maxilla. *AJR*. 2002;179:1077-8.
9. Ayoub KS, Grimer RJ, Carter SR, Mangham DC, Davies AM, Tillman RM.

- Clear cell chondrosarcoma of bone. Sarcoma. 1999;3(2):115-119.
10. Liu C, Xi Y, Li M, Jiao Q, Zhang H, Yang Q, et al. Dedifferentiated chondrosarcoma: Radiological features, prognostic factors and survival statistics in 23 patients. PLoS ONE. 2017;12(3):e0173665.
 11. Kumar A, Jain V K, Bharadwaj M, Arya RK. Ollier disease: Pathogenesis, diagnosis, and management. 2015;38(6):e497-506.
 12. Prokopchuk O, Andres S, Becker K, et al. Maffucci syndrome and neoplasms: a case report and review of the literature. BMC Res Notes. 2016;9:126.

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