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Case Report .

Mesenchymal Chondrosarcoma of the Mandible: A Rare Entity

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ABSTRACT

Mesenchymal chondrosarcomas are rare malignant connective tissue neoplasms that can develop from the both soft and hard tissues. Localization in the jaws are rare and even more in the mandible. They exhibit characteristic biphasic histopathological pattern but misleading clinical and radiographic findings resembling other jaw tumors. Owing to the aggressive nature of the tumor as well as its metastatic and recurrence potential, the prognosis is poor. We report a rare case of Mesenchymal Chondrosarcoma on the left mandibular body in a 54-year-old female patient.

Keywords: Mesenchymal Chondrosarcoma, Mandible, Malignant Tumor.

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INTRODUCTION

Chondrosarcoma (CS) is an uncommon malignant mesenchymal tumor characterized by abnormal generation of the cartilaginous tissue and failure to produce bone tissue.[1-3] CS is a second most common primary malignant tumor of the bone and mesenchymal chondrosarcoma is that rare, bimorphic histological subtype of CS which accounts for only 1% of total CS in the head and neck region.[2,4,5] Mesenchymal chondrosarcoma (MCs) is however common in the head and neck region, particularly the maxillary alveolus amongst jaws. It is seen more often in younger population unlike conventional chondrosarcomas.[2,4,5] The tumor presents a unique feature of aggressive growth, higher tendency of late recurrence, delayed metastasis and poor prognosis.[2-4,6] Only about 50 cases of MCs of the jaw have been reported in the English-language literature.[3,5] They are distinct tumors arising in unicentric or multicentric locations and from both soft and hard tissues.[3,4,6,7] This neoplasm usually appears in second to third decade of life and affects females slightly more frequently than males.[3,6] We present a case of MCs in a 54 year old female located on the left mandibular parasymphysis region.

CASE PRESENTATION

A 54-year-old female reported to the Department of Oral and Maxillofacial Surgery, second affiliated hospital of Jiamusi University on 18th December, 2015 with a mass on left mandibular region for two months. The patient states no significant medical history and that she found a painless mass about a size of "quail egg" initially in the left molar area but has gradually increased past 2 weeks developing discomfort, facial asymmetry and swelling of size about 3×3 cm. Intraoral examination revealed a firm, lobular swelling in the left lower posterior alveolus. The mucosa presented a normal color and the morphology of tongue, tonsil and soft palate was normal without paresthesia. The mouth opening and secretion from salivary gland was also normal. Lymph nodes were not palpable. CT-Scan revealed irregular hyperdense lumps or calcifications within well-defined radiolucent lesion in the left mandibular body. The differential diagnosis was KCOT (Keratocyst Odontogenic Tumour) as it is more common in the mandibular third molar area, ossifying fibroma due to female predominance, osteosarcoma due to its aggressive nature and all of them exhibits ground glass or cotton wool multilobular appearance in radiographs. An incisional biopsy was performed which showed submucosal undifferentiated short spindle cells resembling like hemagiopericytoma along with small area of chondroid matrix. Immunohistochemistry test was positive CD99, bcl-2, CD57 and Ki-67(25%) and microscopically suggested MCs. On preference of patient's family, she was discharged to seek further treatment on a different hospital. The patient returned to our department a year later on 9th December, 2016. She didn't receive any treatment except for antibiotic therapy and the tumor continued to grow with acceleration for past 2 weeks to a size of 8×7×5 cm accompanied by neuralgia along the area supplied by inferior alveolar nerve. On examination, protruded mass over left mandibular area had soft texture with hard base and ill-defined border. The overlying skin of left cheek posed paresthesia, redness and elevated temperature. Intraoral examination showed ulcerated tumor with pus extending along the vestibule of left lower lateral incisor to second molar (32-37). All of the teeth were fallen off in that quadrant except the third molar (38) and left

central incisor (31) which was third degree mobile along with right central and lateral incisors (41,42). The opening of mouth was limited by second degree trismus along with restricted tongue movements. However, the opening of salivary gland duct was normal and cervical lymph nodes were not palpable. The CT Scan at this stage showed osteolytic destruction and swelling of the chin and left mandibular body with irregular shape and uneven density of the soft tissue clumps. The border was ill- defined and cotton-like, patchy and linear tumor about 7.2×5.2×4.2 cm was noted. The rest of CT was negative for lymphadenopathy or metastasis. The treatment planned was surgical resection of the mandible with left total neck dissection and left Pectoralis major myocutaneous flap transfer under general anesthesia.

A single trifurcate incision was designed and the local anesthesia was administered along the lines of the neck crease. The dissection was proceeded with the blunt dissection of the subcutaneous tissue, platysma and the left submandibular gland was removed with protection of the lingual nerve and ligation of the left submandibular gland duct. The radical neck dissection was performed and the cervical lymph nodes were excised from all five levels of lateral neck as well as resection of spinal accessory nerve, sternocleidomastoid muscle and internal jugular vein was done. The lymph nodes were not adhesive, dark red and had well-defined boundaries. After a midline lip incision revealing a mass, extraction of the upper right third molar (18), upper left 1st molar (26), upper left second molar (27) and upper left third molar (28) was done. Then, resection of mandible from the left ramus till right second premolar (45) with 1 cm margin of normal tissues together with the remaining teeth, lower left lateral one third of lip, overlying skin and bilateral sublingual glands. A pectoralis major myocutaneous flap measuring 20×9 cm was raised and traversed into the neck and resected area and then sutured with complete hemostasis. The tracheotomy was performed for tracheal intubation. The specimen of 7×6×5 cm sent for histopathological examination also revealed consistent findings of MCs where majority of tumor cells showed chondrosarcoma-like differentiation yet few transdifferentiate and scattered into osteoblast-like cells. There was no metastasis in the lymph nodes and surrounding soft tissues. Until a follow up of 6 months, the patient is in good health with no obvious complications.

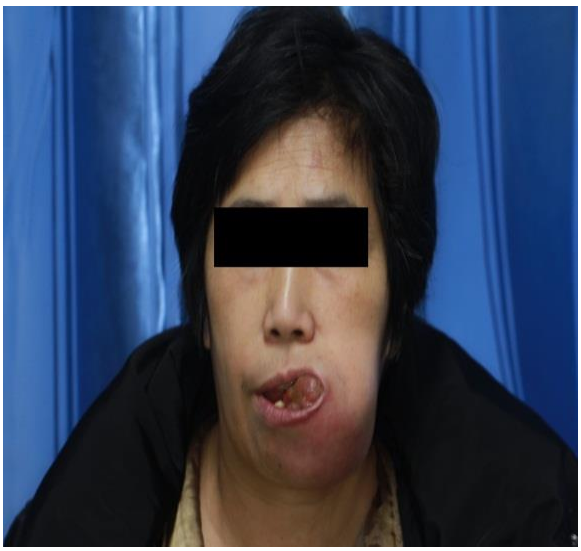

Fig 1: Preoperative photograph of the patient with mesenchymal chondrosarcoma affecting the mandible.

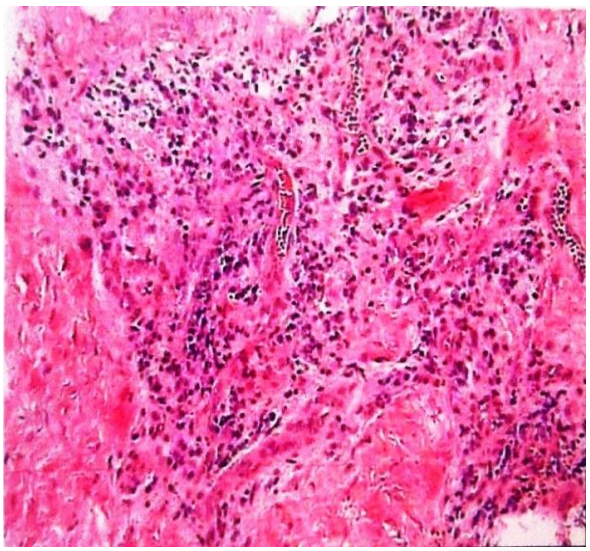

Fig 2: Microscopic section of biopsy tissue showing small undifferentiated spindle cells with arrangement suggesting hemangiopericytoma.

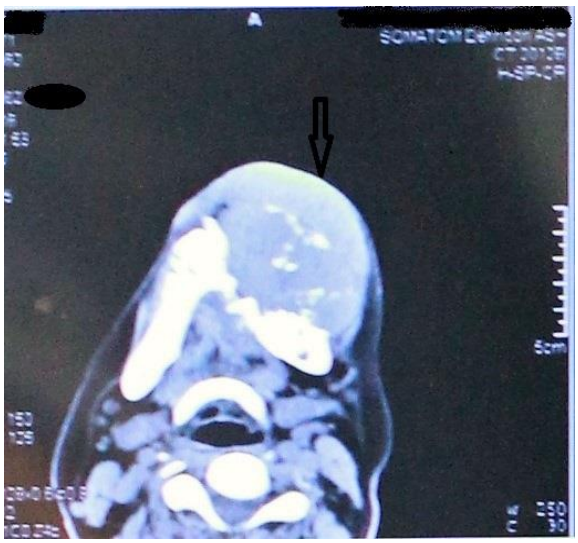

Fig 3: Axial computed tomographic scan of the patient showing extensive involvement of mandible with massive mixed radiolucent and radiopaque lesion.

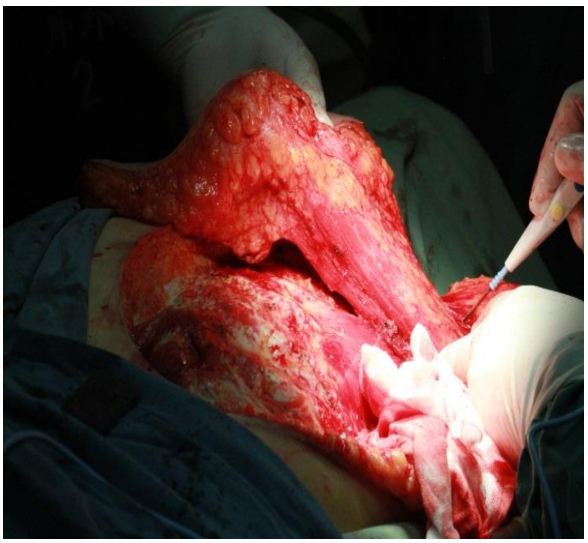

Fig 4: Intraoperative photograph demonstrating elevation of the myocutaneous pectoralis major flap

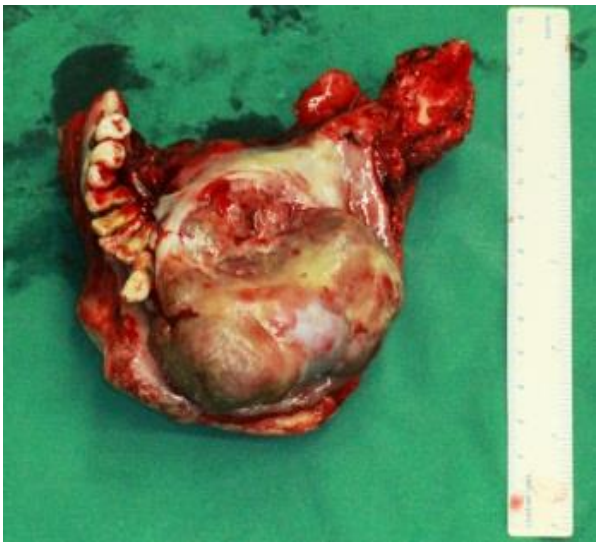

Fig 5: Resected specimen containing mandible, teeth, lateral lower lip and overlying skin.

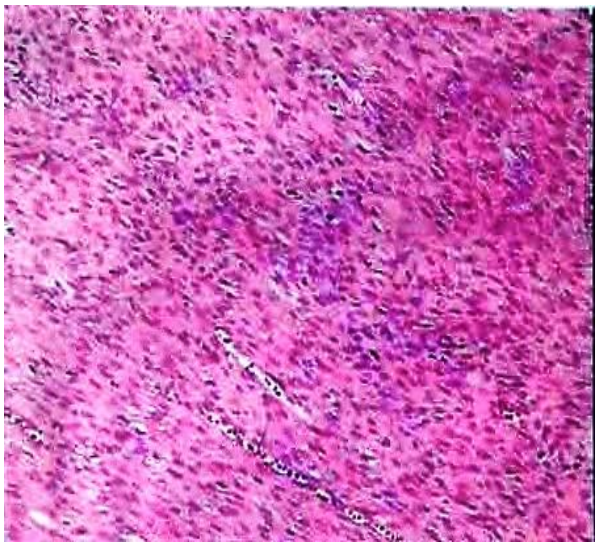

Fig 6: Histopathological section of excised surgical specimen showing cartilaginous islands and transition into osteoblast-like cells.

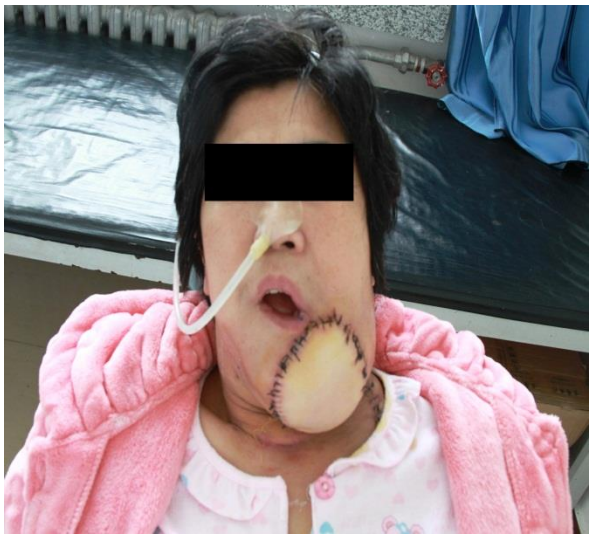

Fig 7: Postoperative photograph of the patient after reconstruction with myocutaneous pectoralis major flap.

DISCUSSION

MCs is a distinctive variant of chondrosarcomas and a rare presentation particularly in mandible.[6,8] It was first documented in 1959 by Lichenstein and Bernstein as a biphasic tumor encompassing spindle cell mesenchyme scattered together with zones of chondroid differentiation.[9-11] According to International Classification of diseases for oncology, chondrosarcomas are classified into 6 subtypes: Not Otherwise Specified, Juxtracortical, Myxoid, Mesenchymal, Clear cell and Dedifferentiated chondrosarcomas.[2,12] CS has also been graded into I, II and III representing low, intermediate and high cellularity, mitotic rate and nuclear size respectively.[2,10] MCs are distinguished by sheets of markedly undifferentiated tiny round or spindle cells with the alternating zones of well differentiated chondroid matrix which are often benign in the presentation.[13] The undifferentiated primitive mesenchymal cells appear similar to anaplastic sarcoma and typically misapprehended with hemangiopericytoma due to its rich vascular component, especially tumors in which the cartilaginous constituent is unremarkable.[6,8,9,13] Other entities often confounded for MCs are osteosarcoma, synovial sarcoma, Ewing's sarcoma, rhabdomyosarcoma, melanoma, osteochondroma.[8,9,13,14] So, it is crucial to examine the multiple sections to confirm a certain diagnosis.[6,8,9,13,15]

MCs arises commonly in osseous than in extraosseous sites.[9] The craniofacial bones, ilium, and ribs are the most common osseous sites.[9] Most MCs of the head and neck region appears principally in the anterior maxilla particularly in the area of pre-existing nasal cartilage followed by the body of the mandible, ramus, coronoid or condylar process, mental symphysis, nasal septum, and paranasal sinuses in a descending order.[10,16] The literature review of MCs of head and neck disclosed female to male ratio of 1.6:1 whereas no gender predilection for MC in the remaining parts of body.[9,16] The MCs of jaw ranges from 5 to 75 years but frequently found in adults between 2nd and 3rd decade of life.[2,9,10,16] In our case, MCs was encountered in body of mandible in female patient of 54-year old.

The most common presentation of MCs of jaws is a painless mass initially but grows rapidly resulting in a facial deformity whereas pain and swelling are the most common presenting symptoms in MCs of other parts of body.[2,6,9,10] It can also be slow-growing, often giving a false impression of a benign tumor.[15] In a review of 41 cases, Takahashi. et al.[8] reported that the average duration of the swelling before the patient presented was about one year. The cortical bone is usually thinned or perforated and occasionally results in pathological fracture. Other symptoms such as epistaxis, gingival bleeding and paresthesia of the lower lip or other facial paresis may occur.[9] Tooth mobility and loss of teeth have also been observed.[15] The clinical presentation was consistent with our current case. Common radiological findings of MCs of the jaw are radiolucency but there may be radiopaque or mixed radiopaque and radiolucent image.[6,9] The current case appeared as osteolytic, radiolucent shadow with a ground glass appearance and foci of calcification. The border of the lesions was also serrated and ill-defined analogous to most MCs of the jaw and this help to differentiate from benign fibro-osseous lesions such as cementifying or ossifying fibromas, which have a similar radiographic characteristic of the ground-glass appearance.[8] However, it is difficult to distinguish between MCs and other cartilaginous neoplasms or osteogenic sarcoma.[6] Hence, an

adequate incisional biopsy is required for making a preoperative diagnosis and immunohistochemical procedure for a specific diagnosis. However, the use of fine-needle aspiration (FNA) in the diagnosis of bone tumors is controversial.[17] The immunohistochemical profile of the present case was also consistent with the literature in that the mesenchymal cells of MC fail to express S100 protein but show positivity for CD99 in almost all cases.[16] In addition, the literature indicates that both types of cells exhibit positive CD57 staining in about two thirds of cases including our current case.[14] In addition, bcl-2 and ki-67(25%) was also positive in our case. Calcification can take place within the chondroid matrix on histological section. Neoplastic cartilage can be substituted by bone in a similar manner to normal endochondral ossification.[10] This might justify the presence of few osteoblast like cells in the histopathological section of the present case. Recent evidences also propose that few portions of terminally differentiated chondrocytes can transdifferentiate to osteoblast-like cells.[18]

The most effective and mainstay therapeutic modality is wide surgical excision, usually with a tumor free margin of 2–3 cm.[1,6,9] The role of preoperative or postoperative radiotherapy and chemotherapy is uncertain till current date.[6,9,11] Huvos et al.[19] subdivided MCs into two sets on the basis of microscopic characteristics , the well differentiated hemangiopericytomatoid and the less differentiated small cell type and informed that the adjunct chemotherapy is less effective for the less differentiated type.[15] Radiotherapy is normally useful in conjunction with the surgery but MCs is a radio resistant tumor.[2,10,16] Hence, radical excision appears to be the most effective treatment for both non-head and neck cases of MCs.[9] However, MCs has a high tendency for recurrence and metastasis as hematogenous spread occurs more frequently than lymphatic metastasis with lung being the most common site of metastasis.[9] Hence, the prognosis is usually poor. The reported 5 year and 10 year survival rate is 48% and 28% respectively.[6,9,10,19]

CONCLUSION

MCs is a unique mesenchymal tumor and relatively uncommon in mandible. Due to the rarity and limited experience to the clinical, radiological and histological features of this entity poses a diagnostic challenge to the clinicians. The review of literature shows that only few cases of MCs in jaws were reported so far. Therefore, we present this rare case of MCs of mandibular body in an attempt to add it in the existing literature. Moreover, it is suggested that the early identification may promote its better prognosis with more aggressive interventions. Long term follow-up and systemic assessment is accentuated for MCs of maxillofacial region.

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