CASE REPORT

Mesenchymal Chondrosarcoma of the Jaws: A Series of two Rare Cases

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ABSTRACT

Chondrosarcoma (CS) is a malignant neoplasm that results in abnormal bone and cartilage growth. Although CS is rare, it is the second most common primary bone malignancy. Mesenchymal chondrosarcoma (MC) is a rare histological variant of CS that accounts for only about 1% of all CS and has high predilection for the head and neck region. It is usually seen in younger age group compared to conventional CS and the maxillary alveolus is the most common site. The tumor is unique because of its aggressive growth with a high tendency for late recurrence and delayed metastasis. We present two cases of MC, one case involving the mandibular condyle and the second, a metastatic CS with the primary in the clavicle.

Keywords: Chondrosarcoma, Mesenchymal chondrosarcoma, Metastasis.

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CASE REPORTS

Case 1

A 22-year-old male patient reported to the dental hospital with the complaint of a swelling on the right side of the face since the past 2 months.

History revealed that the swelling has been progressively growing in size over the period of 2 months. An ulcer developed in the right buccal mucosa about 1 month back which did not heal with medications. The patient had a restricted mouth opening and numbness in the right side of the lower lip since last 15 days. On examination, a diffuse swelling was seen causing facial asymmetry on the right side of the face, ovoid in shape, measuring 7 × 6 cm in size. The swelling was firm in consistency, fixed to the underlying bone and nontender on palpation. No secondary changes of ulcer or sinus were seen and there was no discharge. Intraorally, an ulceroproliferative growth was seen in the right retromolar region extending anteriorly upto 45, measuring about 3 × 3 cm, the ulcer was covered with slough, with irregular margins. It was indurated and tender on palpation. There was no discharge from the ulcer, surrounding area firm in consistency with bicortical expansion of the mandible in the 45 to 47 region. A provisional diagnosis of a malignant bony neoplasm was given (Fig. 1).

Panoramic radiography revealed decreased radiodensity in the right ramus, with erosion of the condylar outline. There was a typical ‘sunburst’ appearance in the condyle extending up to the angle of the mandible on the right side (Fig. 2).

CT scan revealed osteosclerosis and erosion of the condyle extending upto the ramus, a nonenhancing mass with mottled calcifications was seen in the axial sections measuring 6.4 × 6.5 cm. The classic ‘sunburst’ appearance is also seen. The coronal images also show extension of the involvement of the marrow space (Figs 3 and 4).

An incisional biopsy was then done; histological examination revealed highly cellular connective tissue stroma with formation of large amounts of chondroid matrix (Fig. 5). The nuclei of the chondrocytes were large and pleomorphic with an open chromatin pattern (Fig. 6).
There was evidence of cytoplasmic vacuolization and the presence of typical binucleate forms and there numerous mitotic figures with few foci of calcification. Immunohistochemistry was done and the chondrocytes were positive for S100. Histopathology was suggestive of MS. The patient underwent surgery and chemotherapy.

**Case 2**

A 31-year-old male patient reported to the dental hospital, with the chief complaint of swelling and pain in left posterior maxilla for the past 2 months.

History revealed that the swelling was started as a small one and gradually grew to the present size. He also had pain while swallowing. There was the history of a similar painful swelling in the left clavicle 6 months back for which he had undergone surgery (Fig. 8).

On examination, an ulceroproliferative growth was seen in the left maxillary alveolus measuring approximately 6 × 4 cm in size, extending from the left maxillary premolar region into the tuberosity and soft palate (Fig. 7). There were surface lobulations with ulceration posteriorly, covered by a greyish white slough. It was firm in consistency and tender on palpation. The lesion was provisionally diagnosed as a malignant neoplasm.

Panoramic radiography revealed decreased radiodensity in the left maxillary alveolus with destruction of the floor of the maxillary sinus and soft tissue mass in the sinus. There was the evidence of patchy radiopacities in the otherwise osteopenic lesion.

CT scan revealed a 5.3 × 6.2 × 6 cm hypodense region with specks of hyperdense areas, eroding the posterior maxillary wall. Tumor infiltration into the retromaxillary region, pterygoid musculature and lateral wall of the nose was evident. A space occupying lesion seen eroding the lateral and medial walls of the maxillary antrum on the right side and almost filling it. The CT images were suggestive of an aggressive malignant lesion (Figs 9 and 10).
An incisional biopsy was done and histopathology revealed characteristic bimorphic pattern composed of sheets of undifferentiated round and spindle cells along with areas of chondroid matrix of variable cellularity (Fig. 11). Cells of chondroid differentiation exhibited both nuclear and cytoplasmic pleomorphism, hyperchromasia along with numerous abnormal mitotic figures. Areas of ossification were also evident. Immunohistochemistry was done and the chondrocytes were positive for S100 (Fig. 12). Histopathological features were suggestive of MC. Correlating with the surgical history and clinical findings, a final diagnosis of MC of maxilla metastasized from clavicle was made. The patient underwent surgery but had recurrence in the clavicle and later passed away before he could undergo any further treatment.

**DISCUSSION**

Phemister in 1930\(^1\) defined chondrosarcomas as sarcomas of bone containing abundant cartilage. Lichtenstein and Jaffe\(^2\) established the criteria for classification of chondrosarcomas. They defined chondrosarcomas as arising from full-fledged cartilage and never containing osteoid or bone stroma. According to the International Classification of Diseases for Oncology,\(^3\) CS has been classified into six subtypes, on the basis of the tumor location, the histological characteristics of the malignant cartilage cells, and the makeup of the surrounding matrix material associated with the tumor are as follows:

1. Chondrosarcoma NOS (Not otherwise specified)
2. Juxtacortical chondrosarcoma
3. Myxoid chondrosarcoma
4. Mesenchymal chondrosarcoma
5. Clear cell chondrosarcoma
6. Dedifferentiated chondrosarcoma

MC is a rare histologic type of CS that was first described by Lichenstein and Bernstein in 1959 as a biphasic tumor with areas comprising of spindle cell mesenchyme.
interspersed with areas of chondroid differentiation accounting for only about 1% of all chondrosarcomas. It develops from a pluripotential mesenchymal stem cell which can differentiate into angioblastic, fibroblastic and cartilaginous structures.

Approximately one-third of mesenchymal chondrosarcomas are found outside bone within a variety of soft tissues, while two-thirds of these tumors are intraosseous. Most chondrosarcomas of the head and neck region occur in the maxilla; others are seen to occur less commonly in the body of the mandible, the ramus, the nasal septum and the paranasal sinuses. In the maxilla, the most common location is anterior alveolus where preexisting nasal cartilage is present. Both our reported cases are extremely rare as one is in the mandibular ramus with extensive condylar involvement and the other in the posterior maxilla metastasizing from the clavicle.

Chondrosarcomas do not show any gender predilection. The tumor can occur between 10 and 80 years of age. However, it is more common during the second and third decades of life, therefore it is a malignancy of the young individuals as seen in our cases. There are no specific clinical signs or symptoms but the first clinical symptom is a painless swelling or mass, rapidly growing to a large size and leading to facial deformity, nasal obstruction, expansion of the bone and its subsequent perforation as it enlarges, surface ulceration may also be seen. Later symptoms include pain and mobility of the teeth in the region. Rarely facial paralysis can also occur.

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Radiographically, chondrosarcoma is seen as an irregularly shaped, ill-defined radiolucency with randomly scattered, opaque mottled areas. The lesion may appear lobulated and may elevate the adjacent periosteum to produce radiating laminae in a sunray pattern as seen in the first case. Perforation of the cortex of the involved bone by a cartilaginous lesion is considered a sign of probable malignancy. Since these are not pathognomonic signs of chondrosarcoma the radiographic differential diagnosis also includes fibrosarcoma and osteosarcoma. Evaluation of the maxillary region on conventional radiographs is somewhat limited due to anatomical superimposition and two dimensional displays. CT performed with contrast enhancement is the imaging modality of choice because it best delineates the bony involvement while defining local invasion into the adjacent soft tissues as seen in both our reported cases.

Depending on the cellularity, nuclear staining (hyperchromasia) of the tumor cells and size of the nuclei, chondrosarcomas have been graded into three types:

- **Grade I (or low grade):** These tumors are characterized by the presence of benign cartilage, have a relatively uniform and lobular histologic appearance. Presence of atypical cells including binucleate forms may also be recorded.
- **Grade II (or intermediate grade):** These tumors are characterized by a higher cellularity with a greater degree of nuclear atypia, hyperchromasia with often having myxoid stroma and enlarged chondrocyte nuclei.
- **Grade III (or high grade):** These tumors are characterized by a higher cellularity, marked cellular and nuclear pleomorphism, nuclear hyperchromasia and increased mitosis with occasional presence of giant cells.

Histologically, MC has two characteristic components. The first is a dense population of anaplastic small cells either in solid sheets or in a hemangiopericytoma-like pattern and the second is marked by a characteristic chondroid matrix. Both our cases were characteristic of MC as dense
population of anaplastic cells in a chondroid matrix were seen and since there was marked cellular and nuclear pleomorphism, nuclear hyperchromasia; both cases were considered high grade. Immunohistochemistry also can be done for difficult cases, chondroid areas are positive for S100 protein and neurone-specific enolase is focally positive for primitive mesenchymal cells. Both our cases were S100 positive for chondrocytes.

Wide surgical excision is the mainstay of treatment for MC in the jaw bones. These tumors are radioresistant and chemotherapy can be used as an adjuvant therapy after wide surgical excision is made. Our cases were treated with wide surgical excision followed by chemotherapy, but the second case, the primary was in the clavicle which had recurrence and the patient passed away. Generally the prognosis of MC is considered to be poor and the grade, the size of the tumor, the adequacy of tumor resection margins may be prognostic indictors. Local recurrence occurs and may indicate subsequent metastasis; hence adequate treatment and long-term follow-up, including periodic systemic evaluation, are required for patients with MC of the maxillofacial region.

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