Abstract

Chondrosarcoma (CS) is a malignant tumor characterized by the formation of cartilage and usually arises in peripheral long and flat bones. It forms second largest group of bone tumors with less than 10% cases occurring in the head and neck region. It represents 0.1% of total head and neck malignancies.

This is a case report of a 19 year old male patient who reported with a complaint of facial swelling, nasal stuffiness and intraoral swelling with respect to the hard palate. Based on the radiologic and histopathological investigations, the patient was diagnosed with low grade chondrosarcoma reported to be rare in the maxilla.

Keywords:
chondrosarcoma, maxilla, recurrence, metastasis
**Introduction**

Chondrosarcoma (CS) is a malignant tumor characterized by the formation of cartilage and usually arises in peripheral long and flat bones. These are the second largest group of bone tumors, only after osteosarcomas accounting to 26% of all malignancies with 5-10% of the cases occurring in the head and neck region. Pure hyaline cartilage is produced by the tumor cells resulting in cartilaginous growth. It can be a primary lesion but may also develop from preexisting cartilaginous lesions like solitary or multiple osteochondroma. Certain characteristics of slow growth, local recurrence and late metastasis of these tumors makes it imperative to diagnose it in early stage and suggests aggressive treatment. Systematic review shows only few reported cases.

**Case Report**

A 19 year old male patient reported with a chief complaint of nasal stuffiness and repeated nasal discharge since 6 months. Patient presented with a swelling in the nose since 4 months associated with difficulty in breathing and foul smelling nasal discharge, however no bleeding was noted. Patient gave a history of swelling in the oral cavity since 3 months which was followed by a diffuse swelling on the right middle third of the face since 2 months.

On extraoral examination, a diffuse swelling was noted with respect to right middle third of the face, measuring about 4-5 cm in maximum dimension extending superoinferiorly from the infraorbital margin to the right corner of the mouth and anteroposteriorly from ala of the nose to 4 cm posteriorly. No localized rise of temperature was noted. Swelling was non tender on palpation, soft in consistency. Right submandibular lymph nodes were palpable, enlarged, tender, firm and mobile.

Otolaryngeal examination revealed firm, tender pink mass in the right nasal cavity and deviation of the nasal septum towards the left side.

On intraoral clinical examination, a dome shaped well defined solitary swelling was present with respect to the right side of the palate, extending upto the midline. The swelling was approximately 5cm in maximum dimension anteroposteriorly extending from mesial aspect of first premolar to maxillary tuberosity region and mediolaterally from marginal gingiva to 3 cm towards midline (fig: I). On palpation the swelling was firm in consistency with no associated tenderness. No bleeding or discharge on digital pressure was noted. Based on the patient’s history and clinical examination a provisional diagnosis of benign soft tissue tumor involving maxillary sinus and nasal cavity was arrived upon. Various
investigations done were computed tomography, fine needle aspiration cytology and incisional biopsy following which a diagnosis of chondrosarcoma was given.

Computed tomography (CT) (fig:II) showed large, expansile soft tissue attenuation in the nasal cavity with minimal peripheral enhancement on contrast administration. Non expansile soft tissue opacification of right maxillary, right ethmoid and right sphenoid sinus was noted with significant bone remodeling suggestive of neoplastic lesion.

Fine needle aspiration cytology of the swelling in the hard palate was done which on examination revealed cytological features of cartilaginous neoplasm. Histopathological examination after an incisional biopsy from the nasal mass, confirmed the diagnosis of Low grade chondrosarcoma.

Patient was treated with right total maxillectomy for low grade chondrosarcoma (fig: III) followed by post treatment radiotherapy. A post surgical obturator was fabricated for the maxilla and patient is currently on regular follow up.

Discussion

Chondrosarcomas are uncommon malignant neoplasms of the cartilage most commonly found in the long bones and pelvis, with 5-10% of the cases occurring in the head and neck region. Chondrosarcomas however form only 0.1% of the total head and neck malignancies. The common sites of occurrence in head and neck region are the maxilla, mandible, nasal septum, sphenoid sinus and ethmoid sinus. Of the 56 patients in the Mayo series, 41.1% were located in the nasal septum, ethmoid, and sphenoid, 25% in the maxillary sinus, 19.6% in the maxilla, 10.7% in the mandible and 3.6% in the tip of the nose. Temporomandibular joint also presents as an unusual site of occurrence with only 17 cases reported in literature.

Chondrosarcomas usually present in the fifth to seventh decades of life. Relatively young age of patients has been found in review of larger series of craniofacial chondrosarcomas. When all head and neck sites are considered the tumor has a slight male predilection with a ratio of 1.2:1.

Chondrosarcoma can present as a primary or a secondary lesion. Secondary chondrosarcoma can occur in persons with Ollier disease (ie, multiple enchondromas) and Maffucci syndrome (ie, multiple enchondromas associated with soft tissue hemangiomas). Other benign cartilaginous lesions that have been reported, although rarely to result in secondary chondrosarcoma include solitary enchondroma, synovial chondromatosis, and chondromyxoid fibroma.
Chondrosarcoma in head and neck region presents as a painless swelling. It is associated expansion of buccal and lingual plates. Nasal stuffiness, nasal discharge or epistaxis, headache, blurred vision, diplopia may be present depending on the extent of the lesion.  

Chondrosarcomas can be categorized:

I) Based on the Location as:
1. central
2. peripheral
3. juxtacortical

II) Based on the histological features as:
1. mesenchymal
2. clear cell
3. myxoid
4. dedifferentiated

III) Based on the degree of cellular differentiation as:
1. Low grade
2. medium grade
3. high-grade

Due to overlapping complex structures of the craniofacial bones, it is difficult to assess the tumor and associated calcifications on conventional radiographs alone. Therefore computed tomography and magnetic resonance are beneficial to evaluate the bone and the soft tissue characteristics of the tumor and to differentiate it from other conditions.

On conventional radiography it may present as unilocular or multilocular radiolucencies, opacification of air spaces, densely calcified bone mass or presenting as a sunburst appearance. Cortical destruction occurs late in the course of disease and periosteal bone formation is often limited. Computed tomography scan demonstrates characteristic scattered ring-forming calcifications among a hypo dense matrix and often-bony erosions. A Magnetic resonance image helps delineate the full extent of the soft tissue, typically demonstrating low intensity on T1-weighted imaging, high intensity on T2-weighted imaging, and heterogeneous enhancement with gadolinium contrast.

Chondrosarcoma should be differentiated from other benign and malignant tumors, such as squamous cell carcinoma, adenocarcinoma, inverted papilloma, schwannoma, sinonasal melanoma and benign minor salivary gland tumors such as mucoepidermoid carcinoma and adenomatoid cystic carcinoma.

Radiographic differential diagnosis should include chondroma, osteoblastoma, osteochondromas, meningioma, and fibro-osseous lesions.

Histological classification of chondrosarcoma by Evans et al includes

1) Grade 1 or Well differentiated (low grade) chondrosarcoma with small, densely staining nuclei often with multiple nuclei within one lacunae.
2) Grade II or Moderately differentiated chondrosarcoma is characterized by increased cellularity, significant amount of cells having moderately sized nuclei but a low mitotic rate of less than two mitoses per field.

3) Grade III or poorly differentiated chondrosarcoma shows more than two mitoses/HPF, nuclear size generally greater than seen in grade II also include dedifferentiated tumor.13

Chondrosarcomas are graded histologically according to their degree of cellularity, atypia, mitotic activity, nuclear size, and surrounding matrix composition with the pattern varying from lobulated growth like to pronounced cluttering, myxoid changes with occasional calcification and ossification. Lichtenstein and Jaffe established a histopathological diagnostic criteria (table: 1)3

Table 1: Chondrosarcoma criteria by Lichtenstein and Jaffe

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<th>The presence of many cells with plump nuclei</th>
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<th>More than an occasional cell with two such nuclei</th>
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<th>Giant cartilage cells with single or multiple nuclei, or containing chromatin clumps</th>
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Histological differentials of chondrosarcoma should include osteosarcomas and chondroid chordoma. Since chondrosarcoma carries a better prognosis than osteosarcomas, the differential diagnosis between chondrosarcoma and osteosarcoma is probably the most important distinction to consider when dealing with a malignant cartilaginous tumor in the head and neck region. It is also one of the most difficult since chondroid differentiation in osteosarcomas of the jaw is more common than in other sites. Chondrosarcomas with myxoid changes can resemble chondroid chordoma. The distinction can be made as the tumor cells of chondroid chordomas are positive with keratin markers whereas chondrosarcomas are negative.7

Approximately 20% of tumors metastasis, predominantly to the lungs.12 Autopsy occasionally reveals continuous intravascular growth of the tumor and has been known to extend to the right side of the heart or even to the pulmonary arteries.13 Chondrosarcomas progress slowly and metastasize late. Bone scintigraphy examination reveals poor uptake in the chondrosarcoma itself and morbid uptake in the destroyed area of the affected bone. FDG-PET I has been found useful for grading tumors and for evaluating local recurrences and metastasis. A standardized uptake value of 1.3 has been reported as a border value.
that distinguishes between benign and malignant tumors.\textsuperscript{14}

Management of chondrosarcoma involves multimodality protocols where surgery followed by radiotherapy or chemotherapy is preferred depending on the extent of the lesion, involvement of the associated structures, localized or distant metastasis. The chondrosarcoma of the maxilla is classically treated by radical surgery. Radiotherapy or chemotherapy is usually recommended for adjuvant treatment, higher-grade tumors with a poor prognosis in terms of recurrence and metastasis, residual or recurrent disease and palliation\textsuperscript{15}. Unfortunately, to date studies have not shown adjuvant treatments such as chemotherapy or radiotherapy to have any significant impact on patient morbidity or mortality in the majority of isolated primary lesions.\textsuperscript{16}

Local recurrences are quite frequent accounting to 20–60\% of the cases. They can recur at any time ranging from a few months to several years after the initial diagnosis and treatment. The major contributory factors towards recurrence are incomplete resection and tumor grade.\textsuperscript{17}

According to Fu and Perzin prognosis of chondrosarcoma is variable and dependent on mainly three factors: location and extent of the lesion, adequacy of surgical therapy and degree of differentiation of the tumor.\textsuperscript{18}

Chondrosarcoma of the jaws has poor prognosis as compared to that of the long bones, due to the direct extension of the tumor to the base of skull and distant metastasis, however the prognosis is reported to be good for low and intermediate grade chondrosarcoma\textsuperscript{4}. The five-year survival rate of grade I chondrosarcoma is 90\%, 81\% for grade II chondrosarcoma followed by 43\% for grade III chondrosarcoma\textsuperscript{14}. Follow up at regular intervals with repeated investigations may be necessary due to high recurrence rate and distant metastasis. So early detection may be beneficial to enhance the treatment and thus to improve the quality of life of the patient.
References