Clinicopathologic correlation of chondrosarcoma of mandible with a case report

Sanchita Kundu, Mousumi Pal, Ranjan R. Paul

Abstract

Chondrosarcoma is a rare primary malignant neoplasm of the head, neck, oral and maxillofacial regions. The clinicopathological and radiographic findings are usually characteristic; however, not decisive. The neoplasm is usually treated by wide surgical resection because it is traditionally radioresistant. However, radiotherapy is generally advised for high-grade lesions, and chemotherapy has a palliative role. The treatment and management are primarily guided by the histological grades of the neoplasm. Prognosis of jaw lesions is poor as compared to the lesions affecting the long bones of the body, and the cause of death is usually by direct extension in the base of the skull or due to distant metastasis to lungs and other bones. A clinical case of chondrosarcoma, involving the right half of mandible of a 36 year old male patient is discussed herewith, encompassing the entire gamut of clinicopathological, radiological and treatment modalities rendered.

Keywords: Chondrosarcoma, egg-shell crackling, ground-glass, metastasis, mitotic figures, segmental mandibulectomy, sun-burst

Introduction

Chondrosarcoma is an uncommon, slow enlarging malignant neoplasm that originates from primitive cartilage forming mesenchyme, and produces a pure hyaline cartilage that results in abnormal bone and /or cartilage growth.[1,2] It is the second most common primary bone cancer after osteosarcoma.[2] The involvement of the craniofacial region is very rare, and accounts for less than 10% of all chondrosarcomas.[3]

This neoplasm usually grows within a bone or on its surface.[4] It may occur at any age; however, most frequently is found between the 3rd and 6th decades of life. Males are more commonly affected than females. The usual clinical finding is a painless swelling leading to the expansion of the buccal and lingual cortical plates with occasional premature exfoliation of teeth. Pain may be a late stage feature, and regional lymphadenopathy is very rare.[3]

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Conventional radiographic findings are usually not pathognomonic. Single/ multiple radiolucencies with poorly defined borders / bone destruction with associated calcifications or ground-glass/ sun-burst appearance, and uniform widening of the periodontal ligament (PDL) space, may occasionally be present. However, contemporary imaging techniques like computed tomography (CT) scan, magnetic resonance imaging (MRI) and fluorine-18 fluorodeoxyglucose positron emission tomography (FDG PET) are useful to diagnose chondrosarcoma and to differentiate this malignancy from its benign counterpart.[2,3,5,9]

Conventional chondrosarcomas are characterized by varied light microscopic features and are divided into following four histologic grades primarily depending on cellularity, nuclear staining (hyperchromasia) of the tumor cells and size of the nuclei:-[2,5,6,9]

- 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.

Grade I and Grade II chondrosarcomas of the jaws and facial skeleton are best treated with local resection using 1.5 cm margins of bone and soft tissue. Neither chemotherapy nor radiotherapy is indicated as the primary treatment.

However, the Grade III chondrosarcomas are treated with an initial aggressive resection of 3 cm in bone and 2 cm in soft tissue followed by chemotherapy.[8] Distant metastasis is usually rare; however, does occur in high grade, advanced or recurrent cases to the lungs, sternum and vertebrae.[7] The prognosis of chondrosarcoma of the jaws is poor as compared to that of long bones. The cause of death is usually by direct extension into the base of skull and also through distant metastasis to lungs and bones.[5,9]

Based on the above clinico-pathological, radiological and histopathological findings, a case of chondrosarcoma involving the right side of mandible was diagnosed and has been discussed herewith.

Case Report

A 36 year old male patient reported to the Oral and Maxillofacial Pathology Department of Guru Nanak Institute of Dental Sciences and Research, Panihati, Kolkata, with a chief complaint of swelling involving the right side of face for about 8-9 months. Extra orally, there was presence of a medium sized, firm to hard, slightly tender swelling with ill-defined margins involving the right side of mandible. Overlying skin appeared normal [Figure 1]. Intraoral examination revealed the presence of firm, lobular, slightly tender central mandibular swelling in relation to right —premolar-molar region, associated with expansion of both the cortical plates. Egg-shell crackling was evident over

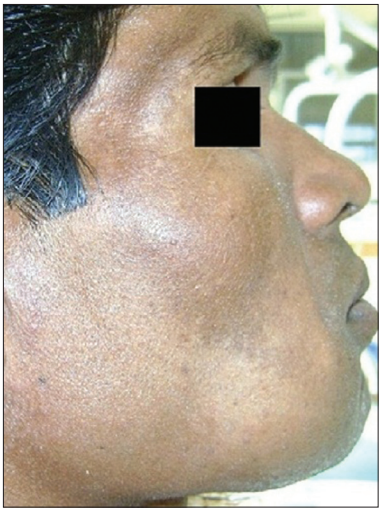

Figure 1: Extra-oral photograph showing the presence of a medium sized diffused swelling involving the right side of mandible, and covered with normal appearing skin of the patient in the case report

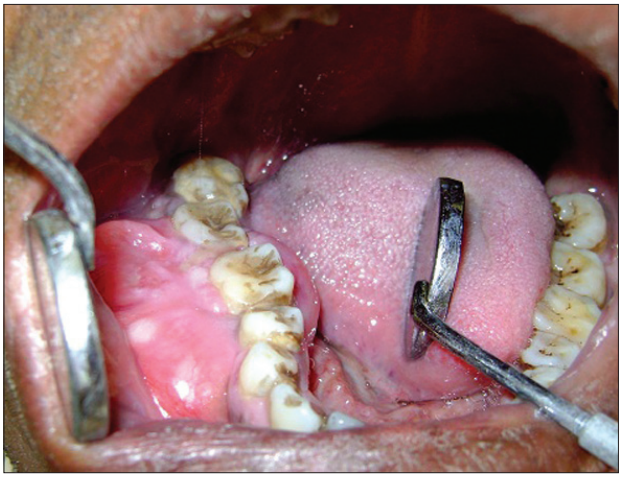

Figure 2: Intra-oral photograph showing the presence of a lobulated central swelling in right mandibular —premolar molar region causing the expansion of both the cortical plates and covered by intact overlying mucosa of the patient in the case report

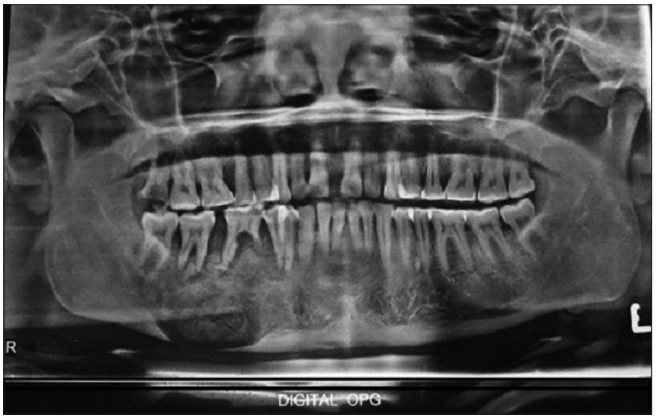

Figure 3: Orthopantomogram (dental panoramic radiograph) showing an irregular radiolucent lesion interspersed with radioopacity with concomitant marked alveolar bone destruction, resorption of the mesial root of first molar tooth and thinning of the lower border of the mandible of the patient in the case report

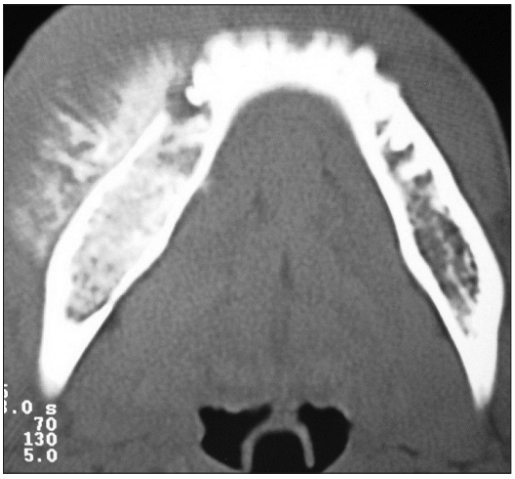

Figure 4: Coronal computed tomography scan revealing the presence of a large osteolytic lesion involving the right body of mandible showing sun-ray appearance and expansion of the cortical plates of the patient in the case report

some areas of the buccal cortical plates. The regional teeth were displaced and mobile while the overlying mucosa had a normal appearance [Figure 2]. Orthopantomogram (dental panoramic radiograph) revealed a large, ill-defined central jaw lesion showing mixed radiographic changes, and characterized by marked alveolar bone destruction, resorption of the mesial root of first molar tooth and thinning of the lower border of the mandible [Figure 3]. Based on the above clinical and radiological findings, an incisional biopsy was performed from the representative site of the lesion. The light microscopic features revealed the presence of a proliferated mass of cartilaginous tissue showing marked cellular and nuclear pleomorphism with nuclear hyperchromatism [Figures 4 and 5].

After considering these microscopic features, a diagnosis of 'High grade Chondrosarcoma' was established and a plain coronal CT scan was advised, which revealed the presence of a large fairly defined osteolytic lesion showing sun-ray appearance and expansion of the cortical plates [Figure 6]. These findings were strongly corroborative to the microscopic diagnosis. The patient was advised to go for surgical treatment. A wide field radical surgery (segmental mandibulectomy with supra-omohyoid block dissection of neck) was performed under general anethesia [Figures 7 and 8]. Post-operative period up to 6 months was uneventful and after that the patient did not report back for further follow-ups at an interval of 6 months. The histopathology of the resected specimen also revealed the characteristic confirmative features of "High Grade Chondrosarcoma."

Discussion

The usual belief is that chondrosarcoma arises from normal chondroid tissues or from embryonic cartilagenous

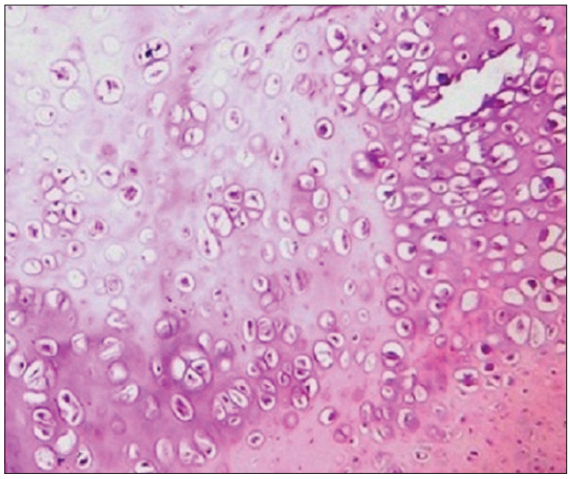

Figure 5: Low power photomicrograph showing proliferated mass of cartilaginous tissue with marked cellular and nuclear pleomorphism of the patient in the case report (H&E, ×10)

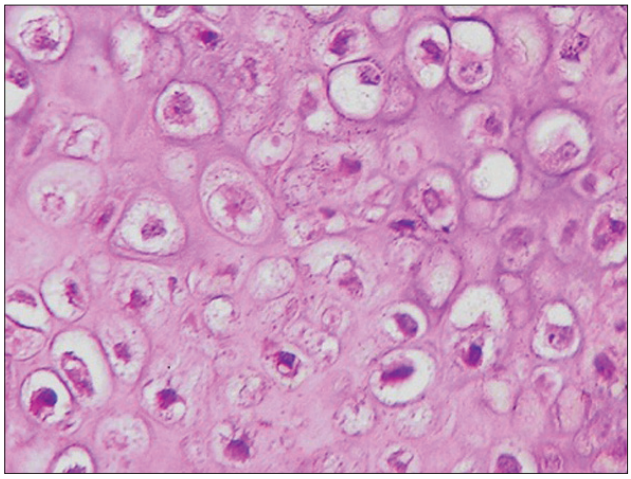

Figure 6: High power photomicrograph showing pronounced features of cellular and nuclear pleomorphism of the patient in the case report (H&E, ×40)

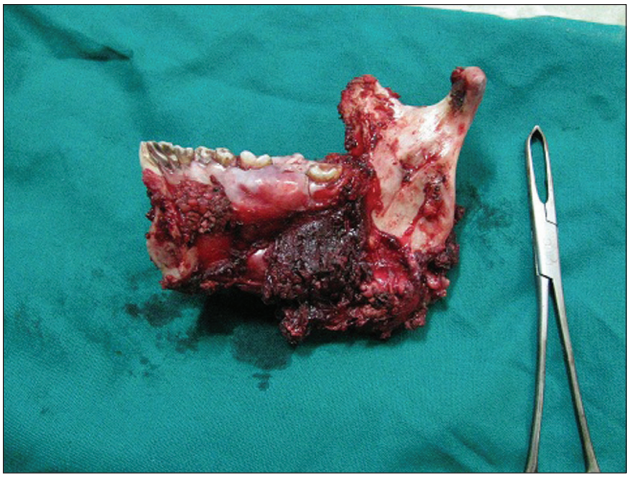

Figure 7: Surgically resected portion of the right half of mandible of the patient in the case report

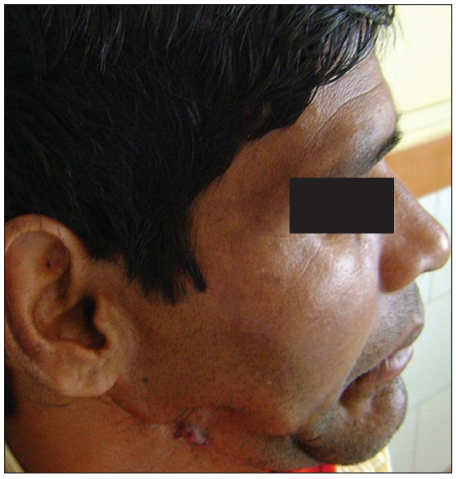

Figure 8: Post-operative extra-oral clinical status of the patient in the case report

rest.[10] However, the exact origin of this malignant neoplasm in the head and neck region is controversial. It may be induced by irradiation, from pre-existing Paget's disease of bone or in association with Fibrous Dysplasia, or from the vestigeal cartilaginous rest.[2] The clinical history of the present case fails to elicit any such associated preexisting disease or lesions. Here, the lesion may have developed from normal chondroid tissue or from the embryonic cartilaginous rest. The patient under discussion was a 36 year old male, having an ill-defined swelling involving the right body of mandible leading to obvious facial asymmetry and the presence of a diffuse, firm, lobular central swelling in the right premolar-molar region intraorally, causing the expansion of both the buccal and lingual cortical plates. No regional lymphadenopathy or neurological abnormalities were recorded. These clinical findings are consistent with the observations reported by the authors of different studies.[3,4]

The conventional radiological findings usually include irregular intramedullary radiolucencies interspersed with punctuate radiopacities, expansion and destruction of the cortical plates, widening of the PDL spaces or even sun-burst appearance at the periphery.[2,3,5] CT scan is superior in defining the peripheral extent of the neoplasm compared to panoramic or flat-plate radiographs.[10] The panoramic view as well as the CT view of the present case was thus very characteristic of chondrosarcoma. The light microscopic features of high-grade chondrosarcoma usually reveal pronounced cellular and nuclear pleomorphism including the occasional presence of giant cells and abundant necrosis.[2,5] The nature of anaplasticity of the tumor tissue and the cells in the present case was strongly mimicking the features of a high-grade chondrosarcoma; however, the giant cells and the areas of necrosis were missing. The entire gamut of discussion involving the clinical imaging and microscopic features of chondrosarcoma thus explains the diagnostic, treatment and management modalities performed in the present case.

Conclusion

Chondrosarcomas are generally radioresistant. High-grade chondrosarcomas commonly do metastasize to regional lymph nodes and to long bones more than do other sarcomas. Survival rate of chondrosarcomas of the jaws in general appear to be poorer than that of chondrosarcomas in other parts of body. Moreover, the response of chondrosarcoma to chemotherapy is much poorer than that of osteosarcoma.

These clinicobiologic characteristics of chondrosarcomas, specially the high-grade ones have rendered them to have a protracted clinical course and high recurrence rates, which in turn stresses on the importance of a proper pre-operative clinicopathological evaluation and a regular periodic post-operative screening.

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