Case Report

Massive dedifferentiated chondrosarcoma affecting whole mandible with high recurrence potential

Niranjan Mishra, Akhilesh Kumar Singh[1], Rahul Agrawal[2], Shreya Singh[3]

Department of Oral and Maxillofacial Surgery, Sriram Chandra Bhanja Dental College and Hospital, Cuttack, Odisha, [1]Departments of Oral and Maxillofacial Surgery, [2]Oral and Maxillofacial Pathology and [3]Orthodontics and Dentofacial Orthopaedics, Institute of Medical Sciences, Banaras Hindu University, Varanasi, Uttar Pradesh, India

Address for correspondence:
Dr. Akhilesh Kumar Singh, Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences, Institute of Medical Sciences, Banaras Hindu University, Varanasi, Uttar Pradesh, India.
E-mail: georgianaks@gmail.com

ABSTRACT

Chondrosarcomas (CS) are slow-growing, malignant mesenchymal tumors characterized by the formation of cartilage by the tumor cells. Benign cartilage-producing tumors within the jaws are extremely uncommon, but most ultimately prove to represent low-grade CS. In the maxillofacial region, the maxilla is more commonly affected than the mandible, it comprises less than 2% of all jaw tumors. Clinically, the tumor presents as a swelling that may be painful and cause loosening of the involved teeth, with widening of the periodontal ligament space. The treatment of such lesions is wide surgical excision with regular follow up. The present report discusses the case of a 55-year-old female patient with massive multiple recurring dedifferentiated CS of mandible.

Key words: Chondrosarcoma, dedifferentiated, jaw tumor, mandible

Introduction

Chondrosarcomas (CS) is a locally aggressive condition resulting in the formation of cartilage rather than bone from the mesenchymal tumor cells. This condition mostly affects femur, humerus, pelvis, and the sacrum accounting for 10-12% of all malignant bone tumors.[1] In the maxillofacial region, maxilla is more commonly affected than the mandible, comprising less than 2% of all jaw tumors. Most CS of facial structures occurs after 30 years of age and the frequency increases with advancing age. There is no racial and gender predilection.[2-4]

CS usually begins with a slow growing mass, with or without pain, associated with displaced and mobile teeth.

Access this article online

Quick Response Code:

Website: www.njms.in

DOI: 10.4103/0975-5950.183874

The areas of occurrence are related to the position of embryonic remnants of cartilaginous tissue such as nasal septal cartilage in the anterior region of the maxilla and Meckel's cartilage in the posterior region of mandible. Approximately 90% of CS are slow growing, low grade, non-metastasizing tumors. The remaining 10% demonstrate aggressive growth, local tissue invasion, and metastasis.[5]

Case Report

A 55-year-old female patient reported to us in March 2009 with complaints of a slowly enlarging mass involving the lower jaw for the last 5 years. The swelling was asymptomatic. She was having difficulty in taking food with multiple malpositioned and mobile teeth. She had the history of a small swelling in the anterior region of the lower jaw that had been curetted 5 years back. Then she had noticed the slowly enlarging mass, which involved the whole of the lower jaw over time. On examination a firm multilobular mass was found with both buccal and lingual cortical expansion, more on the right side than the left side of mandible [Figure 1]. Intraoral examination

showed diffuse swelling obliterating the buccal vestibule with multiple displaced and mobile teeth [Figure 2]. The overlying mucosa was normal and intact. No regional lymphadenopathy was found.

Radiographic study revealed an ill-defined, multilocular radiolucent lesion with massive cortical expansion and multiple displaced teeth ("floating teeth" appearance) [Figure 3a and b]. Computerized tomography (CT) scan showed a large ill-defined homogenous mass measuring 124 × 77 mm involving the whole of the mandible and extending into the infratemporal fossa region, with no intracranial extension [Figure 4].

On incisional biopsy, hematoxylin and eosin (H and E) stained sections revealed multiple islands of cartilage arranged in a lobular pattern, with cells that contained large, plump nuclei and were often binucleated or multinucleated. There was an increase in the number of cells, and each lacuna often contained two or more cells. Areas of hemorrhage and sheets of pleomorphic, hyperchromatic spindle cells were present [Figures 5 and 6]. A diagnosis of dedifferentiated CS was concluded.

Complete resection of the mandible was planned for the patient. Under general anesthesia, tracheostomy was done to prevent postoperative upper airway obstruction, and the tumor was exposed via apron incision over the bilateral submandibular region. Complete resection of the mandible was done and a few parts of the overlying adherent skin was removed [Figures 7-9]. After complete removal of the tumor, hemostasis was achieved, drains were fixed, and wound was closed in three layers. Postoperative recovery was uneventful. She was regular to follow-up till 18 months post-surgery [Figure 10]. After 5 years, she reported to us with massive recurrence over the bilateral face. The firm, diffuse, nontender swelling involving the malar, preauricular, and temporal regions led to a very horrific facial appearance [Figure 11]. A CT scan showed a normal brain, an intensely enhanced soft tissue mass in the masticator space, and infratemporal space on both sides with extension to temporoparietal convexity (extra calvarial) of the left side [Figures 12a and b]. She was terminally ill, with difficulty in taking food, and not interested in further treatment.

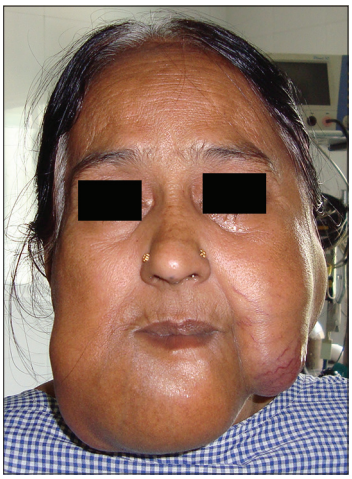

Figure 1: Preoperative photograph shows diffuse multilobular swelling over the bilateral lower face

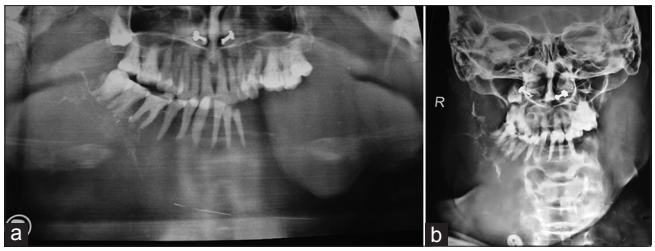

Figure 3: (a and b) Orthopantomogram and posteroanterior view showing an ill-defined, multilocular radiolucent lesion with massive cortical expansion and multiple displaced teeth ("floating teeth" appearance)

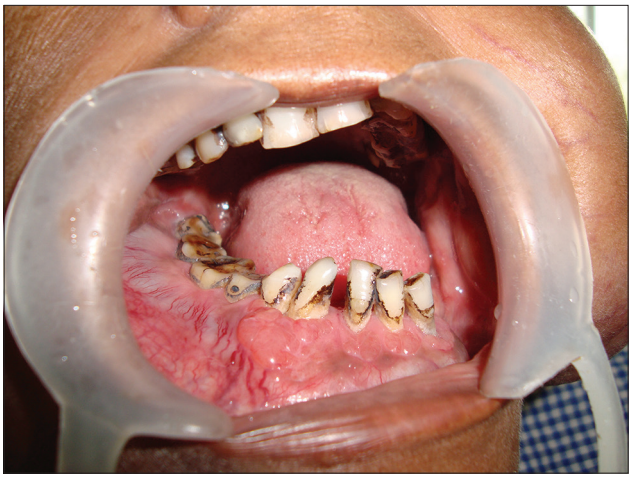

Figure 2: Intraoral examination shows diffuse swelling obliterating the mandibular buccal vestibule with multiple displaced and mobile teeth

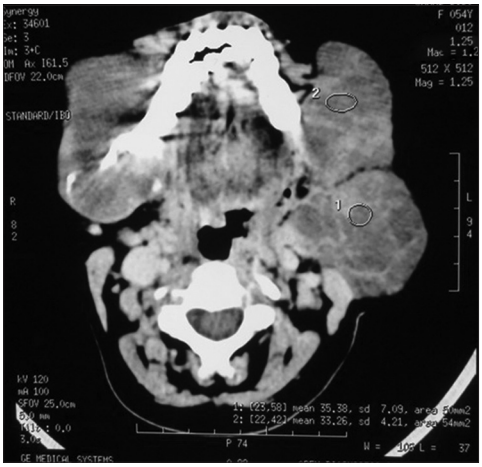

Figure 4: Computed tomographic scan shows a large ill-defined homogeneous-mass involving the whole of the mandible and extending into the infratemporal fossa region, with no intracranial extension

Discussion

CS arises from mesenchymal stem cells and undergoes a partial differentiation to form chondroblastic differentiation and even definable cartilage. In 1942, Lichtenstein and Jaffe described CS as a lesion that develops directly from a sarcomatous stroma, developing

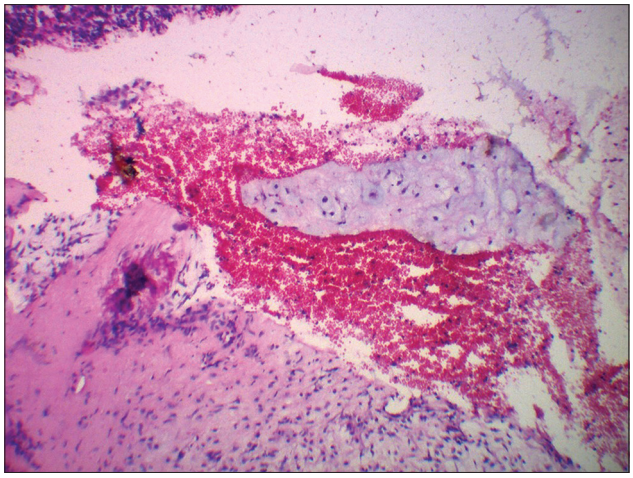

Figure 5: Photomicrograph (H and E; x10) reveals island of cartilage with cells containing large, plump nuclei and sheet of spindle cells

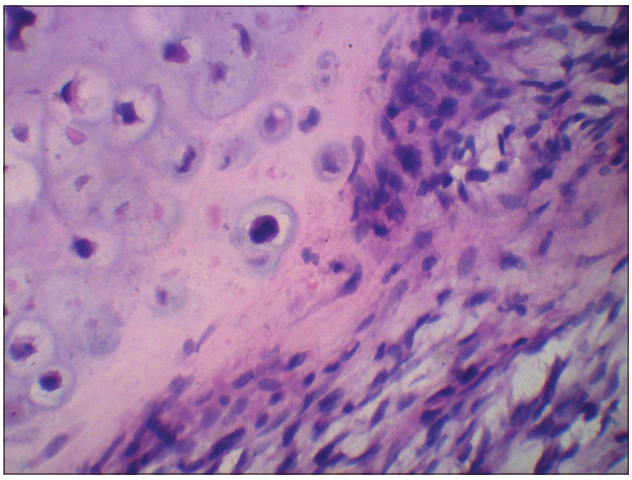

Figure 6: Photomicrograph (H and E; x40) shows increased number of spindle cells with pleomorphism and hyperchromatism, and cartilaginous area having lacunae often containing two or more cells each

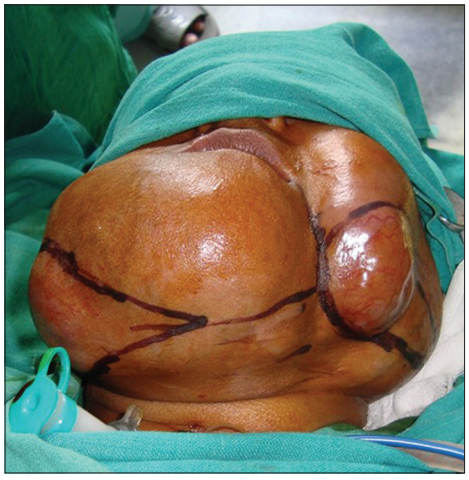

Figure 7: Intraoperative picture shows incision planning

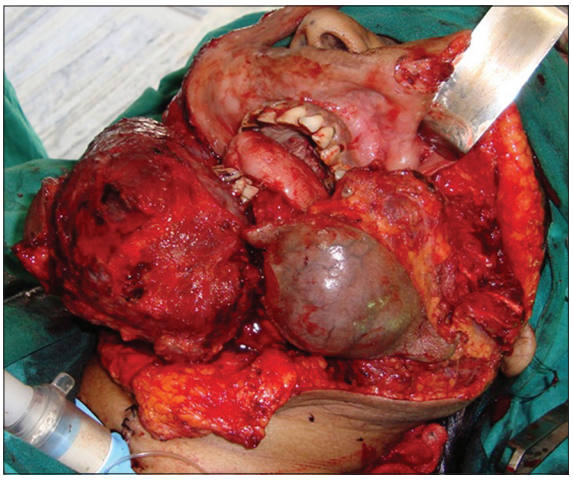

Figure 8: The tumor exposed with apron incision over bilateral submandibular region

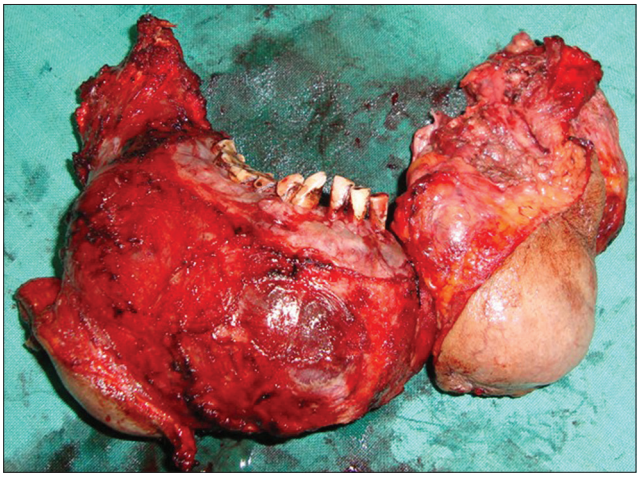

Figure 9: The resected specimen of the involved whole of the mandible

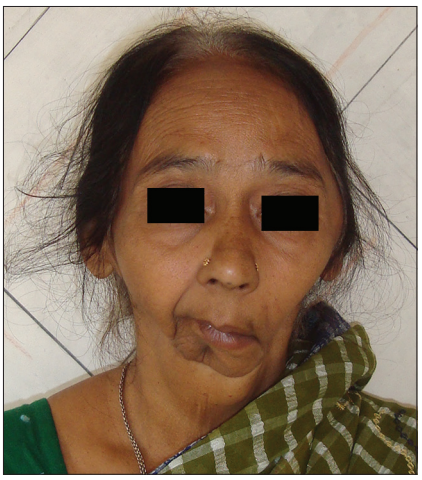

Figure 10: Postsurgery1 year

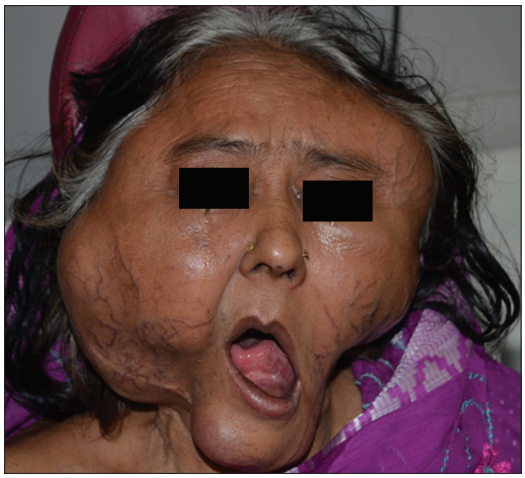

Figure 11: After 2.5 years, massive recurrence over the bilateral face

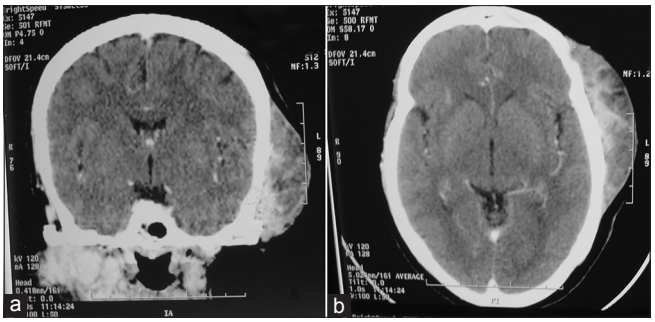

Figure 12: (a and b) Computed tomographic scan with axial and coronal view shows normal brain, intensely enhanced soft tissue mass in the masticator space, and infratemporal space on both sides with extension to temporoparietal convexity (extracalvarial) of the left side

from full-fledged cartilage and not showing neoplastic osteoid tissue and bone.[6] The presentation is that of a slow-growing mass. Pain may or may not be a presenting symptom. The mass will emanate from bone as an irregular lytic lesion that will palpate as a firm-to-hard lobulated soft tissue mass. Teeth involved with the lesion will be displaced and mobile. The mass is only rarely ulcerated. Most CS will be seen either in the anterior part of the maxilla or in the posterior body region of the mandible.[7]

Radiographically it may appear as irregular intramedullary radiolucency causing cortical expansion and destruction. Punctate radiopacities may be present because of dystrophic calcifications or focal ossifications of cartilage.[8] In the tooth-bearing areas, a widening of the periodontal ligament space (Garrington's sign) may be seen as an early sign of CS, just as it is an early sign of osteosarcoma.[4] Because cartilage itself and cartilaginous tumors are well demonstrated by magnetic resonance imaging (MRI), this modality may provide a better delineation of tumor extent than a CT scan. Although metastasis of CS is less frequent than with an osteosarcoma or other sarcomas, a chest radiograph is required to rule out this most likely place for a metastatic focus.

Microscopically it is characterized by the formation of malignant cartilage without deposition of osteoids from a sarcomatous stroma. The cartilage cells have large, plump nuclei and are often binucleated or multinucleated. There is an increase in the number of cells, and lacunae often contain two or more cells. Pleomorphism and hyperchromatism are also present. Histologic grading is important with regard to prognosis in CS. In 1977, Evans *et al*. classified CS into three grades I, II, and III based on cellularity, nuclear size, and mitotic rate. Grade I tumors tend to have a lobular pattern and two or more cells within a lacuna. There is endochondral ossification, and myxoid and cystic areas may develop. Grade II tumors show an increase in cellularity, with retention of lobules and ossification, while grade III tumors are markedly cellular, with a proliferation of spindle cells. The lobular pattern is lost. In the jaws, grade I tumors predominate. Various subtypes were also introduced such as conventional CS, clear cell CS, myxoid CS, mesenchymal CS, and de-differentiated CS.[9] Our case showed features of the de-differentiated variant of grade II CS.

Differential diagnosis based on clinical and radiological features initially suggest a benign odontogenic tumor or a benign tumor of the bone. If the lesion is entirely radiolucent, the clinician may consider an ameloblastoma or odontogenic myxoma. If some punctate radiopacities are identifiable, the lesion will resemble a calcifying epithelial odontogenic tumor, an ossifying fibroma, an immature osteoblastoma, or a cavernous hemangioma of bone. The more obviously aggressive presentations with irregular radiolucencies and perhaps neurosensory loss would be consistent with an intraosseous carcinoma, an osteosarcoma, and a malignant fibrous histiocytoma. A true CS cannot demonstrate bone formation from a malignant mesenchymal stroma. Such entities are actually osteosarcomas, which are especially important, since many osteosarcomas of the jaws and facial bones have significant chondroblastic portions within them. However, if tumorous bone arises from the cartilage rather than from the malignant stroma, it remains a true CS. CS are second to osteosarcomas in their frequency as primary sarcomas of bone. Their overall incidence represents about 25% of all primary sarcomas of bone. However, of these, the face and jaw area represents only 2%.[10]

CS are slow growing tumors with a tendency toward local recurrence after surgery. However, with recurrence they exhibit rapid and aggressive growth. Because of their slow growth and their tendency to undergo neural invasion only later in their course, they are often

mistaken for "benign chondromas" or "cartilaginous rests". Several have presented with previous biopsies identifying "cartilage" where the patient was informed that it merely represented "ectopic cartilage". This false impression of a cartilage hamartoma or a benign cartilage forming tumor is often supported by histopathologic features that will appear to be benign because of mature cartilage with little stroma and mostly a single nucleus in each lacuna.[4,7]

Management of CS is dependent on its grading. The common low-grade CS (grades I and II) of the jaws and facial skeleton are best treated with a local resection using 1.5 cm margins for bone and soft tissue. Neither chemotherapy nor radiotherapy is indicated as primary treatment. These lesions may be invasive but they typically grow slowly; lymph node metastasis is therefore rare, and elective neck dissection is not necessarily required.[11,12] The uncommon high-grade CS (grade III) is treated with an initial aggressive resection of 3 cm in bone and 2 cm in soft tissue followed by chemotherapy. The prognosis for the more common low-grade CS is excellent. The 5-year survival rates are 90% for grade I and 81% for grade II. However, the less common, high-grade (grade III) CS is associated with a 29% 5-year survival rate. High-grade (grade III) CS more often fail to be cure das distant metastasis (66%), most often to the lungs, sternum, and vertebrae have been reported. However, lymph node metastasis also can develop if the initial therapy did not include a neck dissection.[3,9,11]

CS was traditionally regarded as a radio resistant tumor, and radiotherapy was therefore generally reserved for high-grade lesions (as a postoperative adjuvant therapy) and for surgically unresectable lesions.[13] However, Harwood and others reported that CS was radiosensitive and potentially radiocurable.[14] Hence the role of radiotherapy in its management is controversial.

Conclusion

We must state that the appropriate modality in managing CS, whether surgery alone (with or without neck dissection) or adjuvant radiotherapy or chemotherapy, is unclear. Management of CS should depend on its behavior and recurrence potential.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

References

1. Burkey BB, Hoffman HT, Baker SR, Thornton AF, McClatchey KD. Chondrosarcoma of the head and neck. Laryngoscope 1990;100:1301-5.
2. Saito K, Unni KK, Wollan PC, Lund BA. Chondrosarcoma of the jaw and facial bones. Cancer 1995;76:1550-8.
3. Ruark DS, Schlehaider UK, Shah JP. Chondrosarcoma of the head and neck. World J Surg1992;16:1010-6.
4. Garrington GE, Collett WK. Chondrosarcoma. II. Chondrosarcoma of the jaws: Analysis of 37 cases. J Oral Pathol1988;17:12-20.
5. da Rosa Santos OL, Moreira AM, Lopes Cardoso IC, Rodrigues Terra BA, Bellizzi A, Ramos-e-Silva M. Sarcoma of Meckel's cartilage. Int J Dermatol 1996;35:353-4.
6. Lichtenstein L, Jaffe HL. Chondrosarcoma of bone. Am J Pathol 1943;19:553-89.
7. Hackney FL, Aragon SB, Aufdemorte TB, Holt GR, Van Sickels JE. Chondrosarcoma of the jaws: Clinical findings, histopathology, and treatment. Oral Surg Oral Med Oral Pathol 1991;71:139-43.
8. Takahashi K, Sato K, Kanazawa H, Wang XL, Kimura T. Mesenchymal chondrosarcoma of the jaw-Report of a case and review of 41 cases in the literature. Head Neck 1993;15:459-64.
9. Evans HL, Ayala AG, Romsdahl MM. Prognostic factors in chondrosarcoma of bone: A clinicopathologic analysis with emphasis on histologic grading. Cancer 1977;40:818-31.
10. Marx RE, Stern D. Malignant neoplasms of bone. In: Marx RE, Stern D, editors. Oral and Maxillofacial Pathology: A Rationale for Diagnosis and Treatment. 1st ed. Ch. 18. Illinois: Quintessence Publishing Co; 2003.p. 810-4.
11. Murayama S, Suzuki I, Nagase M, Shingaki S, Kawasaki T, Nakajima T, *et al.* Chondrosarcoma of the mandible. Report of case and a survey of 23 cases in the Japanese literature. J Craniomaxillofac Surg 1988;16:287-92.
12. Weiss WW Jr, Bennett JA. Chondrosarcoma: A rare tumor of the jaws. J Oral Maxillofac Surg 1986;44:73-9.
13. McNaney D, Lindberg RD, Ayala AG, Barkley HT Jr, Hussey DH. Fifteen year radiotherapy experience with chondrosarcoma of bone. Int J Radiat Oncol Biol Phys 1982;8:187-90.
14. Harwood AR, Krajbich JI, Fornasier VL. Radiotherapy of chondrosarcoma of bone. Cancer 1980;45:2769-77.

How to cite this article: Mishra N, Singh AK, Agrawal R, Singh S. Massive dedifferentiated chondrosarcoma affecting whole mandible with high recurrence potential. Natl J Maxillofac Surg 2015;6:224-8.

Source of Support: Nil. **Conflict of Interest:** None declared.