

Chondrosarcoma of maxilla

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Abstract

Chondrosarcoma is a rare malignant cartilaginous tumor of the head and neck region. It is a slow-growing tumor and is mostly detected in the anterior maxilla and the base of the skull. We report a case of “Grade II Chondrosarcoma” of the posterior right side of the palate invading maxillary sinus and perforating the floor of the orbit.

Keywords: Carcinoma, maxillary sinus, cartilaginous tumor

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INTRODUCTION

Chondrosarcomas of the head and neck region are rare malignancies, accounting for 1% of chondrosarcomas affecting the whole body.^[1] Chondrosarcomas of the jaws are more common in men, with a male-to-female ratio of 2:1.^[2] On gross examination, the cut surface of chondrosarcoma shows a translucent, blue-gray or white color resembling hyaline cartilage.^[3] There may be areas containing myxoid or mucoid material along with cystic areas.^[3] Histopathologically, chondrosarcomas are hypercellular when compared to an enchondroma wherein the chondrocytes are atypical in appearance with variations in size and shape and contain enlarged hyperchromatic nuclei.^[3] Myxoid changes or chondroid matrix liquefaction is a common feature observed in chondrosarcomas.^[3]

CASE REPORT

A 35-year-old male patient reported to the dental outpatient department of oral and maxillofacial surgery, with the chief complaint of swelling in the posterior region of the right

upper jaw, for a duration of approximately 1 month along with difficulty in breathing on the affected side. No extraoral swelling was appreciated [Figure 1]. Intraoral examination revealed a painless, round swelling measuring 3 cm × 2 cm on the right palatal aspect extending from the region of the second premolar to the second molar [Figure 2]. Overlying mucosa appeared normal. Adjacent dentition was clinically sound and exhibited no signs such as no displacement or mobility. There was a complaint of vision changes. There was no regional lymphadenopathy. On palpation, swelling was soft to firm in consistency, noncompressible, nonfluctuant and tender.

Computed tomography showed a large, well-defined expansile lytic lesion sized 5.3 cm × 4.2 cm × 4.2 cm involving the right maxillary sinus and the corresponding alveolar arch. Inferomedially, destruction of the alveolar arch and hard palate with extension of mass in the oral cavity was observed. Superiorly, the lesion resulted in the erosion of floor of the orbit. On its medial aspect, the tumor mass was extending into the nasal cavity causing erosion and destruction of nasal turbinates. Lateral aspect showed

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perforation of the maxillary sinus [Figure 3]. An incisional biopsy was performed. On gross examination, the resected specimen was soft measuring 1.3 cm × 1.5 cm × 0.9 cm, white in color, varying in consistency (with soft to firm areas) and an irregular surface [Figure 4].

Based on the patient's report and clinical aspect of the lesion, provisional diagnosis considered were pleomorphic adenoma, carcinoma ex pleomorphic adenoma, mucoepidermoid carcinoma, nasopharyngeal carcinoma, chondrosarcoma, osteosarcoma, fibrous dysplasia, squamous cell carcinoma, lymphoma and other malignancies of minor salivary gland origin.

Incisional biopsy was performed, and histopathological examination revealed areas of cartilaginous differentiation with binucleated chondrocytes exhibiting hyperchromatism and pleomorphism. Mesenchymal component was highly

cellular, exhibited pleomorphism and 1–2 mitotic figures per high power field [Figures 5-8]. In view of these findings, the histopathological diagnosis was “Grade II Chondrosarcoma.”

Wide excision with partial maxillectomy and obturator placement was planned. The patient refused for surgical treatment; thus, on a 6-month follow-up, the lesion resulted in proptosis of the affected side along with neurological symptoms. The patient died of distant metastasis within 2 months.

DISCUSSION

The term “Chondrosarcoma” is described as a heterogeneous group of lesion with a diverse morphologic features and clinical behavior.^[3,4] It is a malignant tumor with pure hyaline cartilage differentiation.^[3] It accounts for approximately 20% of malignant bone tumors in



Figure 1: Extraoral profile of the patient is bilaterally symmetrical. No extraoral swelling is seen



Figure 2: Clinical image shows swelling in the posterior region of the right upper jaw extending from the region of the second premolar to the second molar

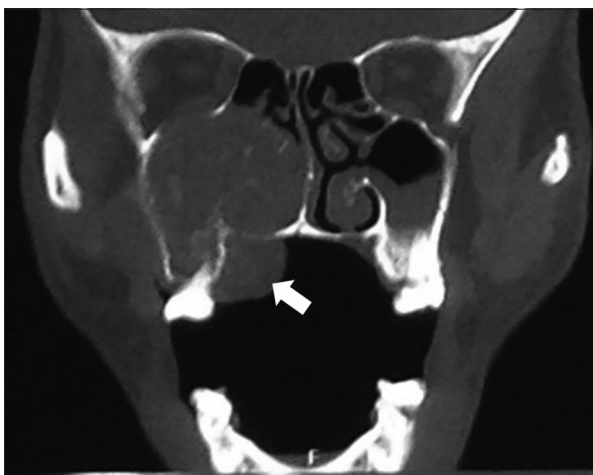


Figure 3: Computed tomography shows a large, well-defined expansile lytic lesion involving the right maxillary sinus and the corresponding alveolar arch (white arrow)



Figure 4: Clinical image shows gross resected specimen, white red in color and an irregular surface

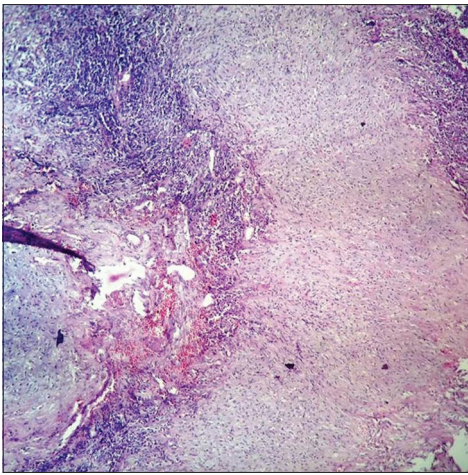


Figure 5: Histopathological examination reveals highly cellular connective tissue stroma at low power view (x4)

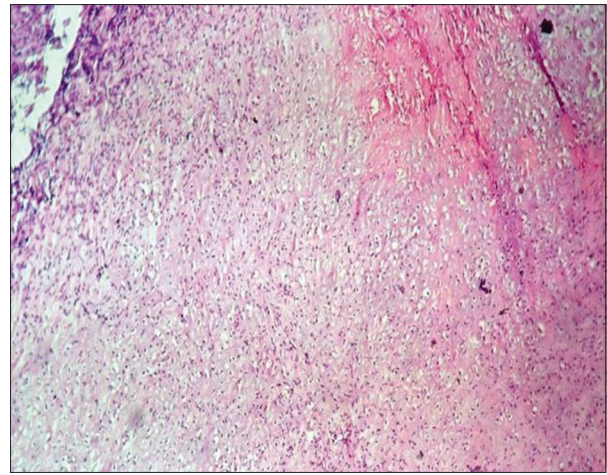


Figure 6: Histopathological examination reveals sheet of cells within lacunae (x10)

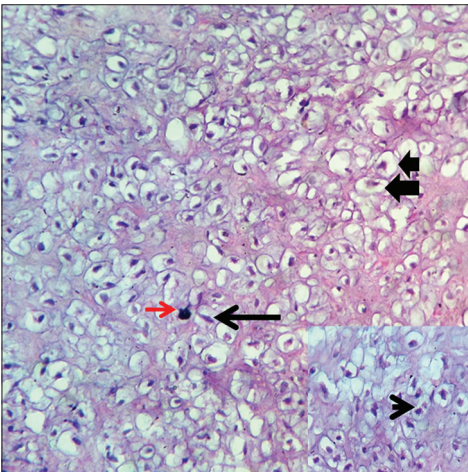


Figure 7: Given H&E stained section shows the presence of increased proliferation of chondrocytes within lacunae with increased nuclear atypia (arrow) nuclear hyperchromatism (red arrow) and cellular atypia (arrowhead), inset showing binucleated chondrocytes (arrowhead) (x40)

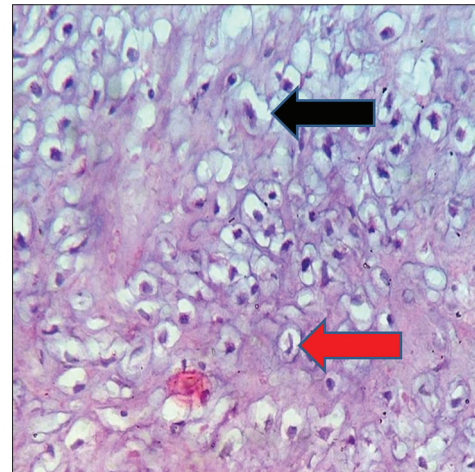


Figure 8: Histopathological examination reveals large abnormal chondrocytes exhibiting nuclear pleomorphism (red arrow) and cellular pleomorphism (black arrow) (x40)

one large series.^[3] More than 90% of chondrosarcoma are primary (conventional) type in the total group of chondrosarcomas.^[3] 1% to 4% of chondrosarcomas affect the head and neck region, mainly the jawbones. In the head and neck cases, the mean patient age ranges from 35 to 45 and most commonly seen in males.^[5] The most common site affected is maxilla than mandible with a proportion of 1.75:1.^[6] It has been postulated that chondrosarcoma originates from remnants of embryonic cartilage precursors from nasal and septal development in the anterior part of the maxilla and from Meckel's cartilage precursors in the posterior aspect of the mandible.^[7] The mucosa is often intact, and it appears as an expanding lesion which is frequently painless.^[3,8] Our case occurred in a male patient with only 35 years as a painless round swelling with intact mucosa. Clinical findings of the present case are

consistent with the observations reported by authors of different studies.

Radiographically, chondrosarcoma presents as a radiolucency with a poorly defined borders showing cortical expansion and destruction.^[7] The radiolucency often contains scattered and variable amounts of radiopaque foci.^[3,7] In the present case, swelling caused destruction of alveolar arch and hard palate with erosion of the floor of the orbit; medially, the tumor mass was extending into the nasal cavity causing erosion and destruction of nasal turbinates.

Microscopically, it shows variable histopathological appearance. The tumor is usually richly cellular with pleomorphic and hyperchromatic chondrocytes, many of which have multiple nuclei.^[9] Several histological parameters are associated with an increased risk of recurrence and

metastasis that include size, location, grade, tumor necrosis, mitotic count and myxoid tumor matrix.^[3,8] Evans *et al.* in 1977 classified chondrosarcomas into Grades I, II and III on the basis of rate of mitotic activity, cellularity and the size of nucleus.^[9] Grade I chondrosarcomas are mildly cellular with an abundant hyaline cartilage matrix, and they rarely metastasize.^[3,9] Occasionally, binucleated cells are seen.^[3] Grade II chondrosarcomas show higher degree of nuclear atypia, particularly toward the periphery of tumor lobules.^[9,10] Grade III chondrosarcomas show greater degree of cellularity, prominent nuclear atypia and mitosis.^[3,8]

In this case, the presence of areas of cartilaginous differentiation with large abnormal chondrocytes exhibiting nuclear atypia, nuclear hyperchromatism, nuclear and cellular pleomorphism within lacunae in a highly cellular connective tissue stroma is strongly mimicking the features of Grade II chondrosarcoma. Binucleated chondrocytes were also seen.

According to WHO, the 5-year survival is 89% for patients with Grade 1 and the combined group of patients with Grade 2 and 3 have a 5-year survival of 53%.^[3] Data gathered from known cases of chondrosarcomas of the jaws indicate that the tumor in this location is exceedingly dangerous and often results in death, either from local invasion or from metastasis to distant sites. To better characterize this tumor, a literature search was conducted in the PubMed database to survey the published case reports of chondrosarcomas in the last many years. Our findings show that chondrosarcomas of maxillofacial region are most commonly seen affecting the maxilla according to Garrington and Collett, 1988,^[11] Smatt, 1977,^[12] Saito *et al.*, 1995,^[6] Mishra, 2008,^[13] Prado *et al.*, 2009^[14] and Gawande *et al.*, 2014.^[15]

CONCLUSION

In the present case, the patient presented all the necessary diagnostic criteria. The present case is unique as a “Grade II Chondrosarcoma” of palatal origin which invaded maxillary sinus and also perforated the floor of the orbit. A thorough lesional examination and a long-term follow-up should be done to ascertain the prognosis.

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.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Izadi K, Lazow SK, Solomon MP, Berger JR. Chondrosarcoma of the anterior mandible. A case report. N Y State Dent J 2000;66:32-4.
2. Ram H, Mohammad S, Singh G, Singh SV. Chondrosarcoma of body of the mandible. Natl J Maxillofac Surg 2013;4:242-4.
3. Bertoni F, Bacchini P, Hogendoorn PC. Chondrosarcoma. In: Fletcher CD, Unni KK, Mertens F, editors. World Health Organization Classification of Tumours. Pathology and Genetics of Tumors of Soft Tissue and Bone. Lyon, France: IARC Press; 2002. p. 247-51.
4. Unni KK, editor. Dahlin's Bone Tumors General Aspects and Data on 11,087 Cases. 5th ed. Philadelphia: Lippincott-Raven; 1996.
5. Panda NK, Jain A, Eshwara Reddy CE. Osteosarcoma and chondrosarcoma of the maxilla. Br J Oral Maxillofac Surg 2003;41:329-33.
6. Saito K, Unni KK, Wollan PC, Lund BA. Chondrosarcoma of the jaw and facial bones. Cancer 1995;76:1550-8.
7. Huang TC, Monsour PA, Chahoud CD. Parosteal osteosarcoma: Report of a case and review of the literature. Aust Dent J 2010;55:86-91.
8. Robert EM, Diane S, editors. Oral and Maxillofacial Pathology: Rationale for Diagnosis and Treatment. Chicago: Quintessence Publishing Co., Inc.; 2003. p. 810.
9. Kundu S, Pal M, Paul RR. Clinicopathologic correlation of chondrosarcoma of mandible with a case report. Contemp Clin Dent 2011;2:390-3.
10. 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.
11. Garrington GE, Collett WK. Chondrosarcoma. II. Chondrosarcoma of the jaws: Analysis of 37 cases. J Oral Pathol 1988;17:12-20.
12. Smatt V. Chondrosarcoma of the upper jaw. Apropos of a clinical case. Rev Stomatol Chir Maxillofac 1977;78:45-51.
13. Mishra A. Chondrosarcoma premaxilla. Indian J Otolaryngol Head Neck Surg 2008;60:259-62.
14. Prado FO, Nishimoto IN, Perez DE, Kowalski LP, Lopes MA. Head and neck chondrosarcoma: Analysis of 16 cases. Br J Oral Maxillofac Surg 2009;47:555-7.
15. Gawande M, Swastika N, Chaudhary M, Patil S. Chondrosarcoma of maxilla. J Oral Maxillofac Pathol 2014;18:423-7.