A Rare Case of Grade II Chondrosarcoma of the Maxilla: Diagnostic Pitfalls in Histopathology

Mohanty Rajat[1], Sangamesh N. C[2], Panda Abikshyeet[3], Pathi Jugajyoti[4], Raghuvanshi Malvika[5]

ABSTRACT

Background

Chondrosarcoma is a rare malignant mesenchymal tumor characterized by persistent cartilaginous differentiation, comprising only 0.1% of head and neck tumors. Within the craniofacial region, commonly affected sites include the larynx, nasal cavity, maxilla, ethmoid and sphenoid bones, and mandible. This report presents a case of Grade II chondrosarcoma of the maxilla presenting a dianostic dilemma in incisional biopsy sample. Case Presentation: A 59-year-old male presented with a one-year history of painless, progressive swelling and discomfort in the right upper jaw. Intraoral examination revealed a well-defined proliferative mass (6 × 4 cm) on the right palate, firm to bony-hard and tender on palpation. Orthopantomogram (OPG) showed an ill-defined radiolucency in the right posterior maxilla extending into the maxillary sinus, with thinning and resorption of the sinus floor and buccal cortex. Histopathological analysis of an incisional biopsy revealed neoplastic chondrocytes within lacunar spaces in a chondroid matrix, with nuclear pleomorphism and hyperchromasia. Although some areas appeared benign, multiple sections confirmed features consistent with Grade II chondrosarcoma. Surgical management included wide local excision via an intraoral infrastructural maxillectomy.

Conclusion

Craniofacial chondrosarcoma presents significant diagnostic challenges due to its resemblance to benign cartilage tumors. Accurate diagnosis requires a multidisciplinary approach incorporating imaging, biopsy, and thorough histopathological evaluation. Early identification is critical to ensure timely and effective treatment. This case highlights the importance of recognizing rare maxillary malignancies and reinforces the need for vigilance in distinguishing benign from malignant cartilage lesions.

Keywords

Chondrosarcoma, Maxilla, Craniofacial malignancy, Cartilaginous tumor, Diagnostic dilemma, Incisional biopsy, Histopathology, Neoplastic chondrocytes, Maxillectomy, Head and neck tumors

INTRODUCTION

Chondrosarcoma – the silent sculptor of cartilage, is a malignant mesenchymal neoplasm marked by sustained cartilaginous differentiation throughout its progression [1]. It predominantly affects the long bones and accounts for approximately 20–30% of all primary malignant bone tumors. However, its occurrence in the head and neck region is uncommon, representing merely 0.1% of all head and neck tumors and between 1% and 12% of all chondrosarcoma cases [2]. Within the craniofacial region, larynx, nasal cavity, maxilla, ethmoid, sphenoid bone and mandible are the most commonly affected sites [3].

1. Professor, Department of Oral and Maxillofacial Surgery, Kalinga Institute of Dental Sciences (KIDS), Kalinga Institute of Industrial Technology (KIIT) Deemed to be University, Bhubaneswar-751024, Odisha, India
2. Professor, Department of Oral Medicine and Radiology, Kalinga Institute of Dental Sciences (KIDS), Kalinga Institute of Industrial Technology (KIIT) Deemed to be University, Bhubaneswar-751024, Odisha, India
3. Professor, Department of Oral and Maxillofacial Pathology and Microbiology, Kalinga Institute of Dental Sciences (KIDS), Kalinga Institute of Industrial Technology (KIIT) Deemed to be University, Bhubaneswar-751024, Odisha, India
4. Professor, Department of Oral and Maxillofacial Surgery, Kalinga Institute of Dental Sciences (KIDS), Kalinga Institute of Industrial Technology (KIIT) Deemed to be University, Bhubaneswar-751024, Odisha, India
5. Professor, Department of Oral and Maxillofacial Pathology and Microbiology, Kalinga Institute of Dental Sciences (KIDS), Kalinga Institute of Industrial Technology (KIIT) Deemed to be University, Bhubaneswar-751024, Odisha, India

Correspondence:

Pathi Jugajyoti, Professor, Oral and Maxillofacial Surgery, Kalinga Institute of Dental Sciences (KIDS), Kalinga Institute of Industrial Technology (KIIT) Deemed to be University, Bhubaneswar-751024, Odisha, India

Bangladesh Journal of Medical Science Vol. 25. Supplementary Issue 2026

DOI: https://doi.org/10.3329/bjms.v25i10.86644

Chondrosarcoma affecting jawbones was first reported by Miles in 1950 [3]. These tumors are thought to originate from mesenchymal remnants of embryonic cartilage—specifically from the nasal septal cartilage in the anterior maxilla and Meckel's cartilage in the posterior mandible. Maxillary chondrosarcomas are exceptionally rare, accounting for only about 5.76% of all head and neck chondrosarcoma cases, with a higher prevalence in adult males, typically between the second and sixth decades of life. The complex anatomy of the maxillofacial region presents significant diagnostic and therapeutic challenges for both pathologists and surgeons [4,5].

Histologically, the tumor may mimic other cartilaginous or myxoid neoplasms, complicating accurate grading and classification, particularly in small or superficial biopsies. For surgeons, the complex anatomy of the maxillofacial region poses difficulties in achieving clear surgical margins without compromising vital structures, which is critical to reducing the risk of recurrence. Therefore, effective management of maxillary chondrosarcoma demands a multidisciplinary approach, integrating precise pathological assessment with meticulous surgical planning [6]. Owing to their aggressive behavior, local recurrence rates can reach up to 50% within two years of treatment, and metastasis may develop years later, highlighting the need for long-term follow-up [5].

This report presents a case of Grade II chondrosarcoma of the maxilla in a 59-year-old patient, highlighting the diagnostic challenges posed by incisional biopsy samples in differentiating benign from malignant lesions to enable accurate treatment planning. It further underscores the crucial role of a multidisciplinary approach in improving diagnosis, clinical management, and optimizing patient outcomes in this anatomically complex region.

CASE REPORT

A 59 years old male patient, reported to the dental OPD with the chief complain of painless progressive swelling and discomfort in right upper jaw region since 1 year. The swelling was initially small and asymptomatic but has gradually increased in size over time and was associated with nasal obstruction, epistaxis and anosmia of the left side over past 2 months. The patient reported no history of trauma or any identifiable triggering factors relevant to the condition.

Intraoral examination revealed a well-defined proliferative growth on the right side of the palate, measuring approximately 6 × 4 cm. The lesion extended antero-posteriorly from the anterior palate in the region of teeth 11 and 21 to the maxillary tuberosity, abutting the mid-palatal raphe medially and extending laterally into the buccal vestibule. The mass appeared irregular and lobulated, with a pink to reddish hue interspersed with white keratotic areas. **[FIGURE- 1]** The overlying mucosa was inflamed. Adjacent teeth exhibited discoloration with notable plaque and calculus deposits; however, no tooth mobility was observed. On palpation, the lesion was tender and exhibited a firm to bony-hard consistency, suggestive of an osseous origin. No active bleeding was elicited upon provocation.

The reconstructed Orthopantomogram (OPG) revealed an ill-defined radiolucency in the right posterior maxilla extending into the maxillary sinus, with thinning and resorption of the sinus floor and buccal cortical plate. Cortical expansion and areas of breach indicated aggressive bone involvement. Complementary 3D CT imaging showed an extensive osteolytic, irregular, lobulated mass involving the right alveolar ridge, hard palate, and maxillary sinus floor, with extension toward the zygomatic bone and infraorbital region, suggestive of advanced local invasion. **[FIGURE - 2]**

Based on the clinical and radiographic evidence of aggressiveness, a provisional diagnosis of a malignant jawbone lesion like osteosarcoma was made. An incisional biopsy performed under local anesthesia revealed neoplastic chondrocytes residing within lacunar spaces in a chondroid matrix, displaying pronounced nuclear pleomorphism and hyperchromasia. Although several regions exhibited benign-appearing chondroid tissue with lacunae containing round, uniform nuclei, few areas after multiple sampling demonstrated histological features consistent with chondrosarcoma. Given the limited size of the biopsy sample, a differential diagnosis of chondroblastic osteosarcoma was also considered. To assess for possible metastasis, imaging studies including chest radiographs, neck and abdominal ultrasonography, and contrast-enhanced CT of the neck were conducted.

Following histopathological confirmation of chondrosarcoma, surgical management was planned under general anesthesia. A wide local excision along with an intraoral approach to infrastructural maxillectomy was performed. The resulting surgical

defect was managed by secondary closure using a medicated gauze pack, stabilized beneath a preoperatively fabricated palatal surgical splint.

Histopathological examination of the specimen revealed a parakeratinized stratified squamous epithelium overlying a fibrovascular connective tissue stroma, which encompassed a lobulated mesenchymal neoplastic proliferation. The lesion was predominantly cartilaginous, with numerous lacunae housing chondrocytes that exhibited marked cellular atypia. Peripheral lobules displayed spindeloid cells, and occasional chondrocytes were binucleated. Focal areas showed the presence of myxoid changes, and calcifications. The overall histological features were consistent with a malignant cartilaginous tumor, confirming the diagnosis of Grade II chondrosarcoma of maxilla with clear margins. **[FIGURE - 3]** The patient remains asymptomatic and has been under regular follow-up for the past six months.

DISCUSSION

Chondrosarcoma originates from mesenchymal stem cells that undergo partial differentiation, leading to chondroblastic transformation and even definable cartilage formation. In 1942, Lichtenstein and Jaffe defined chondrosarcoma as a malignant tumor derived from mature cartilage, characterized by the absence of osteoid or bone formation within its stroma [1]. Chondrosarcomas may be classified as primary, arising de novo, or secondary, developing from pre-existing benign cartilaginous lesions such as enchondromas or osteochondromas [4]. Although the conventional subtype is most frequently encountered in the head and neck region, other histological variants include myxoid, clear cell, juxtacortical mesenchymal, and dedifferentiated chondrosarcomas [1]. Furthermore, they are classified according to Evans' histological grading system (Grade I–III) based on parameters such as cellularity, nuclear differentiation, and nuclear size. These grading systems evaluate similar histopathological criteria, including increased cell density, nuclear pleomorphism, multinucleation, and the presence of mitotic figures. Each feature is assessed using a point-based approach, aiding in the differentiation between low-, intermediate-, and high-grade tumors, which is critical for determining prognosis and guiding treatment strategies [7]. The present case had no prior history of related lesions and was classified as a primary chondrosarcoma, intermediate type.

Diagnosing chondrosarcoma can be particularly challenging because of its overlapping clinical, radiological, and histopathological features with other tumors, often requiring thorough clinical history and a multidisciplinary approach for accurate diagnosis and effective management. This case concerns a 59-year-old male diagnosed with Grade II chondrosarcoma of the maxilla. Epidemiological data suggest a slight male predominance, with a male-to-female ratio of 1.15:1. The average age of affected individuals is approximately 33.14 years, ranging from 2 to 82 years, with males (mean age 33.38 years) tending to be slightly older than females (mean age 31.78 years). Commonly reported clinical manifestations include pain (20.5%), swelling (50.4%), and facial asymmetry (19.6%), typically developing over an average period of one year [5]. In contrast, our patient presented with less common symptoms, including unilateral left-sided nasal obstruction, epistaxis, and anosmia.

Radiographic imaging plays a crucial role in complementing biopsy results by providing detailed information about the lesion's nature, extent, and relationship with surrounding anatomical structures. This information assists in differentiating between benign and malignant lesions, guiding biopsy site selection, and improving diagnostic accuracy [8]. Radiographically, chondrosarcoma present as an irregular intramedullary radiolucent area associated with cortical involvement. Punctate radiopacities can also be observed, resulting from dystrophic calcification or localized ossification within the cartilaginous tissue [9].

The clinical and radiographic appearance of chondrosarcoma can mimic various benign or malignant jaw lesions. Radiolucent lesions may suggest benign odontogenic tumors like ameloblastoma or odontogenic myxoma, while the presence of punctate radiopacities may resemble calcifying epithelial odontogenic tumors, ossifying fibromas, or immature osteoblastomas. More aggressive features—such as irregular radiolucency or neurosensory deficits—may indicate malignancies like intraosseous carcinoma, osteosarcoma, or malignant fibrous histiocytoma. Unlike osteosarcoma, which shows bone formation from malignant mesenchymal stroma, true condrosarcoma forms cartilage-derived bone.

Biopsy of chondrosarcomas can often yield inconclusive or misleading results, potentially leading to under-treatment. In a systematic review of reported cases, conclusive biopsy findings were obtained in

154 cases (90.1%), while a small proportion showed benign cartilage (1.2%), were initially misdiagnosed as chondroma (3.5%), or confirmed as chondroma (5.3%). These diagnostic challenges highlight the limitations of small biopsy samples and underscore the importance of comprehensive histopathological evaluation [5].

Regarding tumor grading, low-grade chondrosarcoma was the most frequently diagnosed subtype, accounting for 46.9% of cases (n = 105), followed closely by high-grade tumors at 45.5% (n = 102), and intermediate-grade tumors comprising 7.6% (n = 17). El-Naggar et al. outlined key micromorphological criteria for the diagnosis of chondrosarcoma, including: (1) abnormal cartilage characterized by nuclear atypia, prominent nucleoli, hyperchromatic nuclei, and infiltration between lamellar bone; (2) hypercellularity; and (3) disordered spacing of chondrocytes. For accurate tumor grading, biopsy sampling should target regions likely to contain high-grade components, such as areas showing endosteal scalloping, soft tissue extension, or diffusely enhancing zones with limited mineralization [10].

Histopathologically, chondrosarcoma must be distinguished from its benign counterpart, chondroma—a tumor composed of mature hyaline cartilage that closely mimics normal cartilage in appearance. Chondromas typically exhibit hypocellular areas with uniformly distributed, bland-appearing chondrocytes embedded in a basophilic matrix. The chondrocytes are characterized by monomorphic cells surrounded by eosinophilic cytoplasm, with typically one cell per lacuna. Notably, features such as cellular pleomorphism, mitotic activity, and binucleation are absent. In contrast, chondrosarcomas demonstrate hypercellularity, nuclear pleomorphism, multinucleated chondrocytes, the presence of mitoses—hallmarks that are critical for distinguishing them from benign chondromas. Due to their indolent nature and delayed neural invasion, they are often misdiagnosed as benign chondromas or ectopic cartilage. Initial biopsies may misleadingly show mature cartilage with minimal stroma and predominantly single-nucleated cells per lacuna, creating a false impression of a benign lesion [11].

Treatment of chondrosarcoma is often complex and tailored to the individual patient, depending on tumor location, grade, and extent of invasion. Surgical resection poses considerable challenges, largely because the tumor is situated close to vital anatomical structures such as blood vessels, nerves and surrounding tissues. While obtaining clear margins is crucial to reduce the risk of recurrence, it often results in significant tissue removal and potential functional deficits. Additionally, the intricate anatomy of the maxilla elevates the likelihood of surgery complications [12,13].

A systematic review of treatment modalities revealed that radical surgery alone with clear margins was the most commonly employed approach (n = 126, 56.3%). Other treatment strategies included conservative surgery alone (10.3%), chemotherapy alone (1.3%), and radiotherapy alone (2.2%). Combined modalities were also reported: radical surgery with radiotherapy (13.4%), radical surgery with chemotherapy (4.5%), chemotherapy with radiotherapy (1.3%), conservative surgery with adjuvant radiotherapy (4.9%), and a multimodal approach involving radical surgery, chemotherapy, and radiotherapy (5.8%). While chondrosarcomas typically exhibit limited sensitivity to radiation compared to other malignancies, adjuvant radiotherapy can play a valuable role in achieving local disease control, particularly in cases with positive postoperative margins or large tumor burden. These findings underscore the importance of individualized, multidisciplinary management in optimizing outcomes for patients with chondrosarcoma. Thirteen patients underwent neck dissection alongside their primary treatment. Among them, six (46.2%) had regional lymph nodal metastasis, predominantly in high-grade tumors, underscoring the risk of regional spread in advanced cases [5,14].

Local recurrence rates may be as high as 50% within the first two years after treatment. Additionally, metastasis could occur years after the initial diagnosis, highlighting the imperative need for prolonged and vigilant follow-up [15]. The present case has shown no evidence of recurrence or metastasis to date and continues to be under regular follow-up.

CONCLUSION

Epidemiological analysis of rare lesions such as chondrosarcoma is critical, as it contributes to improved diagnostic precision and enables clinicians and pathologists to make informed decisions, ultimately refining treatment strategies and enhancing patient outcomes. The definitive diagnosis of chondrosarcoma remains among the most challenging in tumor pathology due to its close histological resemblance to benign cartilaginous lesions. Therefore, accurate diagnosis

requires a combination of imaging, biopsy, thorough histopathological evaluation, and a multidisciplinary approach. This case underscores the diagnostic dilemma in distinguishing benign from malignant cartilage tumors and highlights the importance of early and accurate identification through histopathological examination to ensure appropriate and effective clinical management.

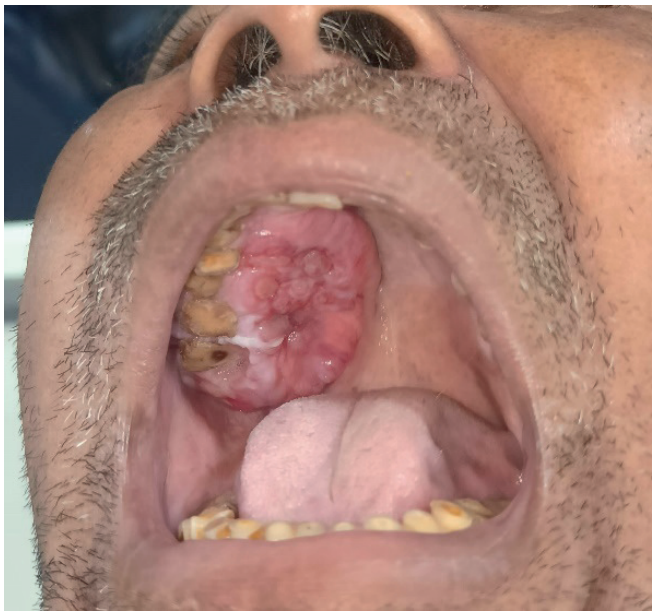

FIGURE- 1 - Intra Oral Presentation

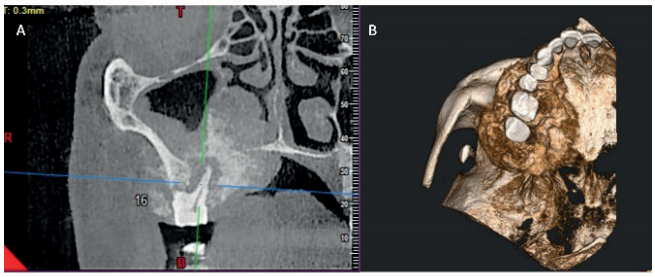

FIGURE- 2 - CBCT Image (A) with 3D Reconstruction (B).

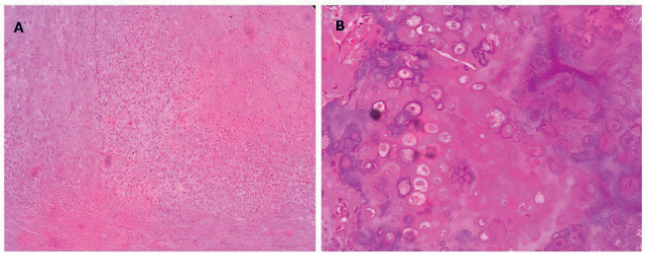

FIGURE- 3 - Histopathology Showing Marked Cellularity with atypia (A) and Atypical Chondrocytic presentation (B).

REFERENCES

1. Gnepp DR. Diagnostic surgical pathology of the head and neck. 2nd ed. Philadelphia: Saunders Elsevier; 2009.
2. Lyoubi M, Oukessou Y, El Krimi Z, et al. Aggressive Mesenchymal chondrosarcoma of the maxilla: Case report. Int J Surg Case Rep. 2022;91:106696.
3. Garrington GE, Collett WK. Chondrosarcoma. II. Chondrosarcoma of the jaws: analysis of 37 cases. J Oral Pathol. 1988; 17(1):12–20.
4. Ruark DS, Schlehaider UK, Shah JP. Chondrosarcomas of the head and neck. World J Surg. 1992;16(5):1010-15.
5. de Souza LL, Pontes FSC, Fonseca FP, da Mata Rezende DS, Vasconcelos VCS, Pontes HAR. Chondrosarcoma of the jaw bones: a review of 224 cases reported to date and an analysis of prognostic factors. Int J Oral Maxillofac Surg. 2019;48(4):452-460.
6. Ragnarsson E, Olafsson SH. Prosthetic restoration following removal of chondrosarcoma in the premaxillary region: a case presentation and follow-up for 16 years. Int J Prosthodont 2004;17:291–6.
7. Evans HL, Ayala AG, Romsdahl MM. Prognostic factors in chondrosarcoma of bone: a clinicopathologic analysis with emphasis on histologic grading. Cancer. 1977; 40(2):818–31.
8. Arlen M, Tollefsen HR, Huvos AG, Marcove RC. Chondrosarcoma of the head and neck. Am J Surg 1970;120:456–60.
9. Hu H, Xu X, Zeng W, Deng H, Yun D, Li G. Low-to moderate-grade myxoid chondrosarcoma in the craniofacial region: CT and MRI findings in 13 cases. Oral Radiol 2015;31:81–9.
10. El-Naggar AK, Chan JK, Grandis JR, Takata T, Slootweg PJ. World Health Organization classification of head and neck tumours. Lyon, France: IARC Press; 2017.
11. 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. pp. 810–4.
12. Kim WS, Jittreetat T, Nam W, Sannikorn P, Choi EC, Koh YW. Reconstruction of the segmental mandibular defect using a retroauricular or modified face-lift incision with an intraoral approach in head and neck cancer. Acta Otolaryngol 2015;135:500–6.
13. Osbon DB, Feinberg SE, Finkelstein MW, Bumsted RM, Zeitler DL. Delayed mandibular reconstruction following removal of a mesenchymal chondrosarcoma: report of a case. Oral Surg Oral Med Oral Pathol 1985;59:557–64.
14. Weber DC, Murray F, Combescure C, Calugaru V, Alapetite C, Albertini F, et al. Long term outcome of skull-base chondrosarcoma patients treated with high-dose proton therapy with or without conventional radiation therapy. Radiother Oncol 2018;129:520-6.
15. Ragnarsson E, Olafsson SH. Prosthetic restoration following removal of chondrosarcoma in the premaxillary region: a case presentation and follow-up for 16 years. Int J Prosthodont 2004;17:291–6.