Case Series

Management of Chondrosarcoma of the Mandible

https://doi.org/10.47210/bjohns.2024.v32i3.128

Krishnappa R,[1] Rahul Shah,[2] Arshad Ali,[1] Jaykumar Patel[1]

ABSTRACT

Introduction

Head and neck chondrosarcomas are rare, typically affecting the anterior maxilla, sinonasal areas, and mandibular molar region. This study analyzes the clinicopathological characteristics of these tumors through three case presentations.

Case Series

This retrospective study included patients diagnosed with chondrosarcoma of the mandible who underwent tumor excision with subsequent reconstruction. All patients presented with rapidly progressing painless facial swelling, and biopsy revealed neoplasms arranged in lobules composed of spindle cells exhibiting moderate cytoplasm, pleomorphic vesicular nuclei, and prominent nucleoli demonstrating moderate nuclear atypia. Central chondroid matrix containing atypical chondrocytes in lacunae was observed, along with a mitotic rate of 7-8/10 hpf, areas of necrosis, calcification, and Ki-67 positivity.

Discussion

Early detection followed by radical surgical resection remains pivotal for achieving a favorable prognosis in mandibular chondrosarcoma. Given the high risk of recurrence, diligent long-term follow-up is imperative.

Keywords

Chondrosarcoma; Mandible; Head and Neck; Oral; Management

According to the World Health Organization (WHO), chondrosarcoma is classified as a malignant tumor characterized by pure hyaline cartilage differentiation, wherein tumor cells form cartilage without bone.[1] Head and neck chondrosarcoma is uncommon, comprising 5% to 12% of all chondrosarcoma cases. While virtually any craniofacial skeletal location can harbor this neoplasm, excluding the larynx, it predominantly manifests in the anterior maxilla and sinonasal structures.[2,3] The molar region of the mandible is reported as the most frequent site for mandibular chondrosarcoma.[4] However, the literature contains few published cases to date. At a tertiary care center, patients with chondrosarcoma of the mandible can benefit from access to advanced diagnostic and therapeutic modalities, as well as expertise from a multidisciplinary team experienced in managing complex head and neck malignancies. Through a coordinated and comprehensive approach, the aim is to achieve optimal oncological outcomes while preserving quality of life for the patient. This study seeks to analyze the clinicopathological characteristics of head and neck chondrosarcomas through the presentation of a series comprising three cases.

1 - Department of Head and Neck Surgery, Kidwai Memorial Institute of Oncology
2 - Department of Head and Neck Oncology, Gujarat Cancer & Research Institute, Ahmedabad

Corresponding author:
Dr. Jaykumar Patel
email: dr.jaypatel@yahoo.com

Case Series

Three female patients, aged between 30 to 40 years, were diagnosed with chondrosarcoma of the mandible. They all presented with painless and rapidly progressive swelling over the face, persisting for 45 to 60 days. (Fig. 1)

Among them, two patients exhibited right-side disease while one patient had left-sided involvement. The tumors

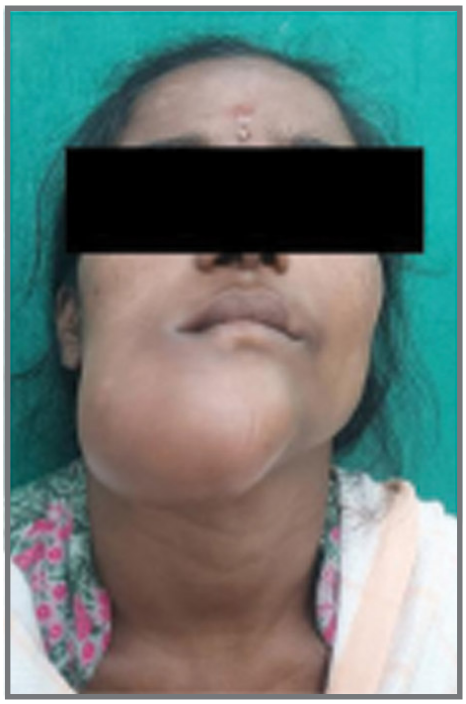

Fig. 1. External Swelling

displayed an ulcero-infiltrative pattern, originating from the mandibular bone and extending to involve the entire half of the mandible along with the overlying skin. Additionally, all patients experienced loosening of teeth in the affected areas. (Figure 2)

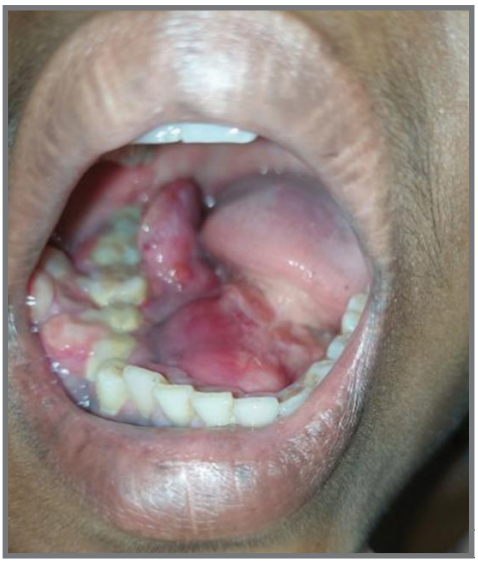

Fig. 2. Intraoral Tumour

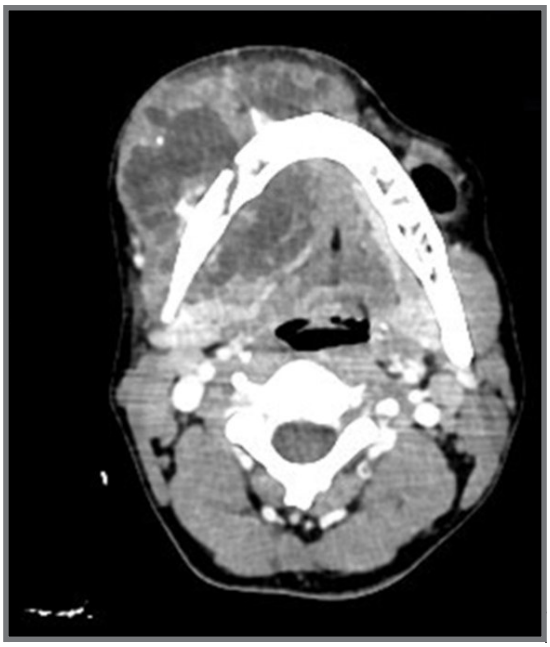

Fig. 3. Computed Tomography Scan

Contrast-enhanced computed tomography revealed erosion of the mandible. (Figure 3)

Nodal metastasis was absent in all cases. One patient underwent PET-CT, indicating an SUVmax of 25.3 for the primary lesion. Histopathological examination of biopsy specimens from two patients exhibited stratified squamous epithelium-lined skin, with deeper dermis revealing a lobulated neoplasm comprising a cartilaginous matrix with atypical chondrocytes in lacunae. Binucleated cells were observed, and spindle cells with nuclear atypia and myxoid stroma were noted at the periphery of the lobules. Mitotic activity ranged from 3-4/10 hpf. Immunohistochemistry showed Ki 67-10%, consistent with Grade I chondrosarcoma. The third patient exhibited similar findings, with higher mitotic activity (7-8/10 hpf) and areas of necrosis and calcification, indicative of grade III chondrosarcoma. Immunohistochemical staining was positive for CD99 and S-100. All patients underwent clinical assessment and were planned for surgical resection. Wide-local excision of lesions, including segmental mandibulectomy with involved skin, was performed in all cases. Reconstruction was undertaken using a free fibula osseo-cutaneous flap in one patient and a pectoralis major myocutaneous flap in the remaining

two patients. Final histopathological reports confirmed chondrosarcoma with free surgical margins. Adjuvant treatment was not administered to any patient. A minimum follow-up of 14 months revealed no signs of residual disease or recurrence.

Discussion

Chondrosarcoma, a malignancy characterized by a cartilage matrix, typically arises in facial bones, comprising 1–3% of all chondrosarcomas.[3] It tends to occur slightly more frequently in males, with a male-to-female ratio of 1.15:1. However, our case series notably featured exclusively female patients, with a mean age of 33 years (range: 2–82 years). The primary complaint, shared among our patients, was painless facial swelling, rapidly progressing over time.[3,4] While the literature commonly cites the maxilla as the most prevalent site for chondrosarcoma, our cases exclusively originated from the mandible.[5] Radiographically, chondrosarcomas often present as osteolytic lesions with radiolucent shadows and irregular borders. Advanced cases may exhibit a characteristic cloud-like matrix with calcified "whorls and arcs," accompanied by features such as endosteal scalloping, cortical disruption, periostitis, or soft tissue mass effect. Similarly, all cases in our series demonstrated gross mandibular destruction.[6] Chondrosarcomas typically manifest as large tumors exceeding 4 cm in size, characterized by a firm, mucoid, or gelatinous consistency with lobulated borders.[7,8] Evans' grading system stratifies chondrosarcomas based on cell density, nuclear size, staining, and mitosis. Grade I tumors display diverse lobular shapes with abundant hyaline cartilage matrix and low cellularity, while Grade II tumors exhibit increased cellularity and less chondroid matrix. Grade III tumors demonstrate high cellularity, marked nuclear atypia, pleomorphism, and extensive necrosis.[9] Differential diagnosis may include chondroblastic osteosarcoma, but the absence of osteoid and neoplastic bone helps differentiate chondrosarcoma. Notably, some chondrosarcomas may exhibit rapid growth following biopsy, emphasizing the need for prompt treatment post-biopsy. In light of a recent case within our patient cohort, where an individual experienced advancement of the condition following incision biopsy from the overlying skin at an external treatment facility, it underscores the importance of vigilance when encountering middle-aged patients presenting with rapidly progressing mandibular swelling, particularly in the absence of tobacco use. Such circumstances warrant a heightened suspicion for chondrosarcoma, given the potential implications of inadvertent involvement of surrounding tissues. Surgical resection with a wide margin (approximately 2-3 cm) remains the optimal treatment approach for mandibular chondrosarcomas. Chondrosarcoma typically displays radio and chemo resistance, rendering them ineffective. Neck dissection is generally unnecessary due to the tumor's low propensity for nodal metastasis.[10] However, a high-grade and recurrent tumor may lead to distant metastasis, commonly affecting the lungs, vertebrae, and sternum. Reconstructive options post-surgery may involve bone grafting with skin or soft tissue grafts. The five-year survival rates vary between grade I (approximately 90%) and combined grade II and III (around 50%) chondrosarcomas. Despite the relatively favorable prognosis, recurrence remains a concern, as observed in previous studies.[11] Fortunately, our patients did not develop recurrence or metastasis during the one-year follow-up period.

References

1. Fletcher CDM, Unni KK, Mertens F. World Health Organization classification of tumors. Pathology and genetics of tumors of soft tissue and bone. Lyon: IARC; 2002
2. Koch BB, Karnell LH, Hoffman HT, Apostolakis LW, Robinson RA, Zhen W, Mench HR. National Cancer Database report on chondrosarcoma of the head and neck. Head Neck 2000;22:408–25
3. Lacerda L, Pontes C, Felipe Paiva Fonseca, Rezende D, César V, Rebelo A. Chondrosarcoma of the jaw bones: a review of 224 cases reported to date and an analysis of prognostic factors. International Journal of Oral and Maxillofacial Surgery. 2019 Apr 1;48(4):452–60
4. Murayama S, Suzuki I, Nagase M, Shingaki S, Kawasaki T, Nakajima T, *et al*. Chondrosarcoma of the mandible. Report of cases and a survey of 23 cases in the Japanese literature. J Craniomaxillofac Surg 1988;16:287 92

5. Shirato T, Onizawa K, Yamagata K, Yusa H, Iijima T, Yoshida H. Chondrosarcoma of the mandibular symphysis. Oral Oncol Extra. 2006;42(6):247–50. https:// doi. org/ 10. 1016/j. one. 2006. 04. 002
6. Sarin V, Singh K, Kakkar V, Singh G. Chondrosarcoma of mandible: A rare case report. Iran J Otorhinolaryngol [Internet]. 2021;33(119):389–94. Available from: http://dx.doi.org/10.22038/IJORL.2021.47251.2553
7. Murphey MD, Walker EA, Wilson AJ, Kransdorf MJ, Temple HT, Gannon FH. From the archives of the AFIP: imaging of primary chondrosarcoma: radiologic-pathologic correlation. Radiographics. 2003 Sep;23(5):1245-78
8. Santini-Araujo E, Kalil RK, Bertoni F, Park YK, editors. Tumors and tumor-like lesions of bone. Springer Nature; 2020 May 5
9. WHO Classification of Tumours Editorial Board. Soft tissue and bone tumors, vol. 3. 5th ed. WHO Classification of Tumours. Lyon: Lyon International Agency for Research on Cancer; 2020
10. Saini R, Abd Razak NH, Rahman SA, Samsudin AR. Chondrosarcoma of the mandible: a case report. Journal of the Canadian Dental Association. 2007 Mar 1;73(2)
11. Prado FO, Nishimoto IN, da Cruz Perez DE, Kowalski LP, Lopes MA. Head and neck chondrosarcoma: analysis of 16 cases. British Journal of Oral and Maxillofacial Surgery. 2009 Oct 1;47(7):555-7.