Radiology Case Reports 20 (2025) 2521–2526

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Maxillary aggressive chondrosarcoma: A rare and challenging case ☆

Fadila Kouhen[a,b,c,*], *Malak Chahid*[a,b], *Reyzane El Mjabber*[a,b], *Othmane Bensalah*[a,b], *Meryem Naciri*[a,b], *Hanae El Gouach*[a,b,c], *Othmane Kaanouch*[b,d], *Najwa Benslima*[a,e], *Rajaa Karmi*[a], *Youssef Mahdi*[f], *Basma El Khannoussi*[a,g], *Mohammed Afif*[h]

[a] *Mohammed VI University of Sciences and Health (UM6SS), Casablanca, Morocco*
[b] *Radiotherapy Department, International University Sheikh Khalifa Hospital, Casablanca, Morocco*
[c] *Laboratory of Neurooncology, Oncogenetic and Personalized Medicine, Mohammed VI University of Sciences and Health (UM6SS), Casablanca, Morocco*
[d] *Hassan First University of Settat, High Institute of Health Sciences, Laboratory of Sciences and Health Technologies, Settat, Morocco*
[e] *Radiology Departmenent, International University Mohammed VI Hospital, Casablanca, Morocco*
[f] *Pathology Department, National Institute of Oncology, Rabat, Morocco*
[g] *Pathology Department, International University Sheikh Khalifa Hospital, Casablanca, Morocco*
[h] *Department of Radiotherapy, Faculty of Medicine of Tangier, Abdelmalek Essaadi University, Tangier, Morocco*

ARTICLE INFO

Article history:
Received 11 January 2025
Revised 25 January 2025
Accepted 4 February 2025

Keywords:
Chondrosarcoma
Maxilla
Radiotherapy
Multidisciplinary management

ABSTRACT

Chondrosarcomas represent 20%-30% of primary malignant bone tumors, but only about 1% occur in the head and neck region. Maxillary chondrosarcomas constitute a mere 5.76% of head and neck cases, predominantly affecting adults between the second and sixth decades of life. Symptoms, including facial swelling, pain, and nasal obstruction, often lead to delayed diagnoses and increased tumor aggressiveness. This case report details a 45-year-old Moroccan male with an aggressive maxillary chondrosarcoma presenting as a progressive left cheek swelling accompanied by significant pain and vision impairment. Imaging studies revealed a large, lytic maxillary lesion, and biopsy confirmed a low-grade chondrosarcoma. Given the tumor's unresectability, the patient underwent radiotherapy, receiving a total dose of 70 Gy with modest reduction in tumor size but ultimately developed metastatic pulmonary lesions, leading to palliative chemotherapy. Despite aggressive management, the patient succumbed after the fourth cycle of chemotherapy. This case underscores the importance of timely diagnosis and multidisciplinary collaboration in managing maxillary chondrosarcomas, highlighting the challenges posed by their aggressive nature and the need for long-term surveillance to monitor for recurrence and metastasis.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: fadila10m@hotmail.com (F. Kouhen).

https://doi.org/10.1016/j.radcr.2025.02.003

Introduction

Chondrosarcomas account for approximately 20%-30% of all primary malignant bone tumors; however, in the head and neck region, they constitute only about 1% of chondrosarcomas overall [1]. Specifically, maxillary chondrosarcomas represent a small subset, comprising around 5.76% of all head and neck cases. Typically arising in adults, these tumors demonstrate a slight male predominance and are most commonly diagnosed between the second and sixth decades of life. Furthermore, their nonspecific symptoms, such as facial swelling, pain, and nasal obstruction, often lead to delayed diagnosis, allowing the tumors to grow larger and become more aggressive [2].

Due to their aggressive nature, local recurrence rates can be as high as 50% within 2 years following treatment. In addition, metastasis may occur many years after the initial diagnosis, underscoring the critical importance of long-term surveillance. Management of maxillary chondrosarcoma usually involves surgical resection, with adjuvant therapies tailored to each individual case.

However, surgical resection presents significant challenges, primarily due to the tumor's location near critical anatomical structures, including nerves, blood vessels, and adjacent tissues [3].

Achieving clear margins is essential for minimizing recurrence; nonetheless, this can lead to substantial tissue loss and functional impairment. Moreover, the complex anatomy of the maxilla increases the risk of surgical complications.

Conversely, radiotherapy serves as a valuable complementary approach, particularly for patients with unresectable tumors or those at high risk for local recurrence. Although chondrosarcomas generally show lower responsiveness to radiation compared to other tumor types, adjuvant radiotherapy can effectively help control local disease, especially in cases with positive margins or larger tumor sizes. Consequently, the application of radiotherapy is often individualized, highlighting the need for multidisciplinary collaboration in treatment planning to optimize patient outcomes.

In this paper, we report an aggressive chondrosarcoma case in a 45-year-old Moroccan patient, emphasizing the challenges associated with diagnosis and treatment planning. Furthermore, we discuss the critical importance of collaboration among specialists to optimize patient outcomes in such complex cases.

Case report

A 45-year-old male presented with a progressive swelling in the left cheek region over the past 6 months. Initially small, the swelling gradually increased in size, leading to noticeable facial asymmetry. The patient reported significant associated pain, nasal obstruction, and vision impairment in the left eye. He experienced difficulty chewing and mild discomfort when pressing on the area. His medical history was unremarkable, with no prior trauma, surgeries, or systemic illnesses, and he denied any recent infections or weight loss.

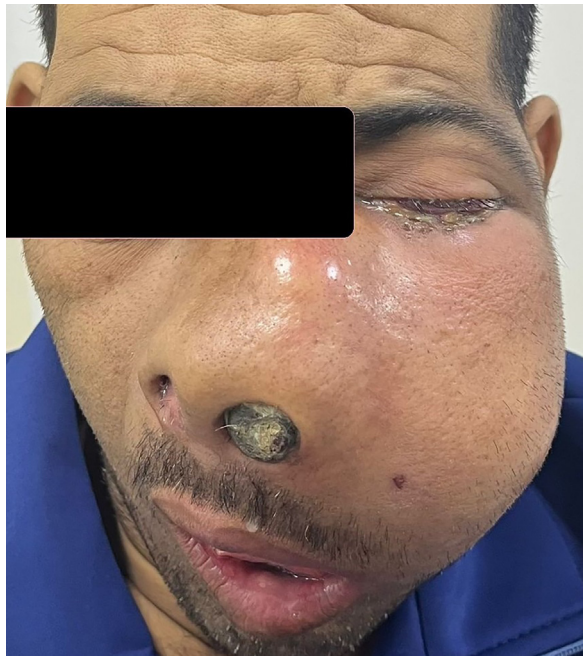

Fig. 1 – Left maxillary swelling with involvement of the left nostril and inflammatory signs in the adjacent regions.

On clinical examination, the left maxillary region revealed a large, firm, nontender swelling measuring approximately 15 cm in diameter (Fig. 1). The swelling was immobile and poorly defined, with no distinct borders, indicating aggressive involvement of surrounding tissues. There were inflammatory signs, such as mild erythema and swelling in the adjacent soft tissues, contributing to the patient's nasal obstruction and difficulty breathing through the left nostril. Intraoral examination showed slight displacement of the upper molar teeth, but there were no signs of mucosal ulceration. The oral cavity appeared otherwise healthy, and there were no palpable lymph nodes in the cervical region. Neurological examination revealed no cranial nerve deficits; however, the patient reported complete loss of vision in the left eye, suggesting potential involvement of the optic nerve or surrounding structures due to the lesion's size and aggressive nature.

Facial CT revealed a large, poorly circumscribed soft tissue mass with irregular margins, centered on the left maxillary sinus. The lesion exhibited extensive infiltration into the adjacent ethmoid and sphenoid sinuses, with invasion of the orbital cavity, including involvement of the extraocular muscles. There was significant osteolysis affecting the bony structures of the left hemiface and the skull base, indicative of aggressive osseous destruction (Fig. 2).

MRI revealed a large, poorly defined mass with irregular contours centered on the left maxillary sinus. The lesion exhibited hypointensity on T1-weighted sequences and heterogeneous hyperintensity on T2-weighted sequences, with diffusion restriction characterized by hyperintensity on DWI and low ADC values. Postgadolinium sequences demonstrated heterogeneous enhancement. The mass extended superiorly into the left ethmoid-maxillary region and sphenoid sinus, encroaching on the left nasal cavity with significant narrowing.

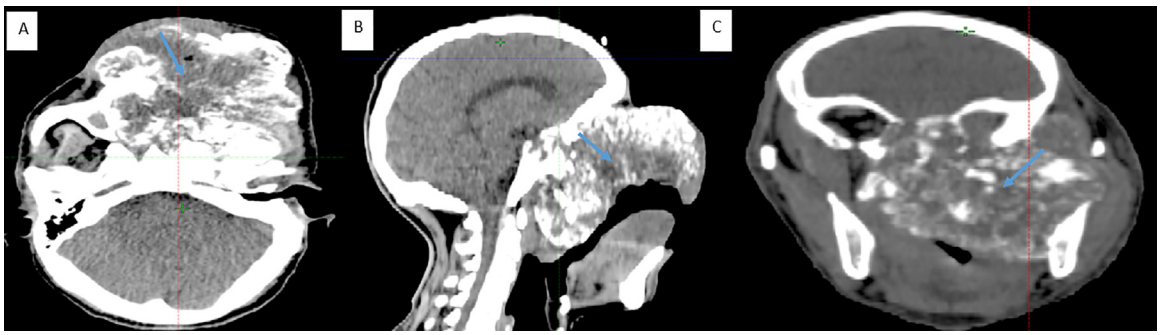

Fig. 2 – Axial (A), sagittal (B), and coronal (C) facial CT images demonstrating a large, Ill-defined invasive soft tissue mass centered on the left maxillary sinus with extensive infiltration into adjacent sinuses, orbital structures, and significant osteolysis of the left hemiface (blue arrow).

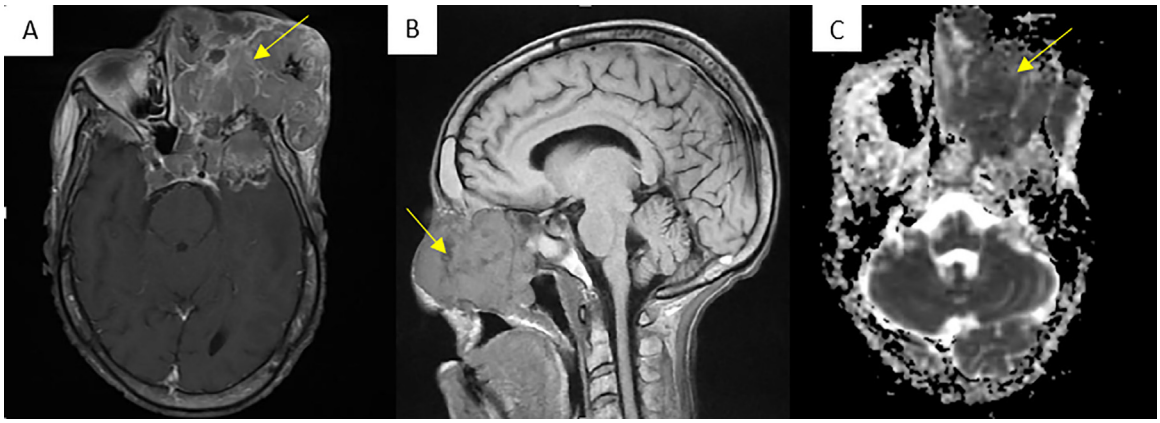

Fig. 3 – Axial MRI (T1-weighted with contrast [A] and diffusion-weighted imaging [C]) and sagittal section (B) Demonstrating an invasive mass centered on the left maxillary sinus with extensive soft tissue and Osseous Involvement (yellow arrow).

Extensive osteolysis of the left hemiface was observed, indicating aggressive osseous invasion, with involvement of the orbital structures (Fig. 3).

A biopsy of the lesion was performed using an incisional approach, allowing for adequate tissue sampling. Histological examination revealed malignant chondroid cells characterized by varying degrees of cellularity. The tumor displayed significant nuclear pleomorphism, with a range of cell sizes and shapes, including atypical nuclei. The presence of myxoid changes and a cartilaginous matrix was noted, which is typical for chondrosarcomas. Additionally, there were scattered mitotic figures, although the overall mitotic activity was low, consistent with a low-grade tumor (Fig. 4).

Immunohistochemical staining was conducted, showing positive expression of S100 protein and collagen type II, further supporting the diagnosis of chondroid origin. The surrounding stroma exhibited moderate fibrosis, and there were no significant signs of necrosis or high-grade features. These findings collectively confirmed the diagnosis of low-grade chondrosarcoma, indicating a malignant cartilage-forming tumor with a relatively favorable prognosis compared to higher-grade variants.

The lesion was deemed unresectable, and the patient was referred for exclusive radiotherapy. During the simulation process, the patient was positioned supine on the treatment couch and secured using a thermoplastic immobilization mask, along with additional devices like headrests, to ensure consistent positioning throughout both the imaging procedure and daily radiotherapy sessions. CT scans were acquired with a slice thickness of 2-3 mm, ensuring comprehensive coverage of the area of interest from the skull base to the carina.

The patient received adjuvant radiotherapy utilizing a 6 MV photon beam delivered by a Truebeam linear accelerator (Varian, CA) through the Volumetric Modulated Arc Therapy (VMAT) technique (Fig. 5). Daily imaging guidance was provided via cone-beam computed tomography (CBCT). A total prescribed dose of 70 Gy was administered over the course of treatment, with daily fractions of 2 Gy, 5 days per week. The Gross Tumor Volume (GTV) included the visible tumor identified on preoperative MRI T1 sequences. The Clinical Target Volume (CTV) was delineated by adding a uniform margin of 1.5 cm around the GTV, while the Planning Target Volume (PTV) was established by extending the CTV with an additional 0.5 cm margin.

The organs at risk (OAR) identified in this treatment plan included the right globe, right optic nerve, optic chiasm, parotid glands, brainstem, spinal cord, and mandible. How-

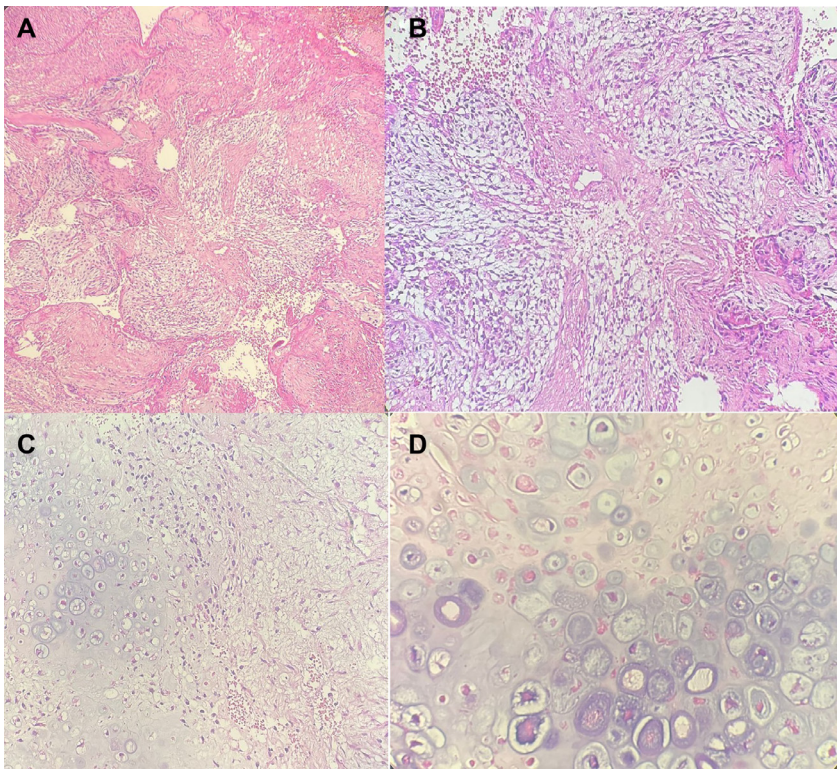

Fig. 4 – Representative micrograph of the tumor. Tumor proliferation presents diffuse growth (A). Myxoid changes (B) and chondroid matrix (C) are observed. Tumor cells presents moderate nuclear atypia and binucleation (D) (Hematoxylin-eosin, x100).

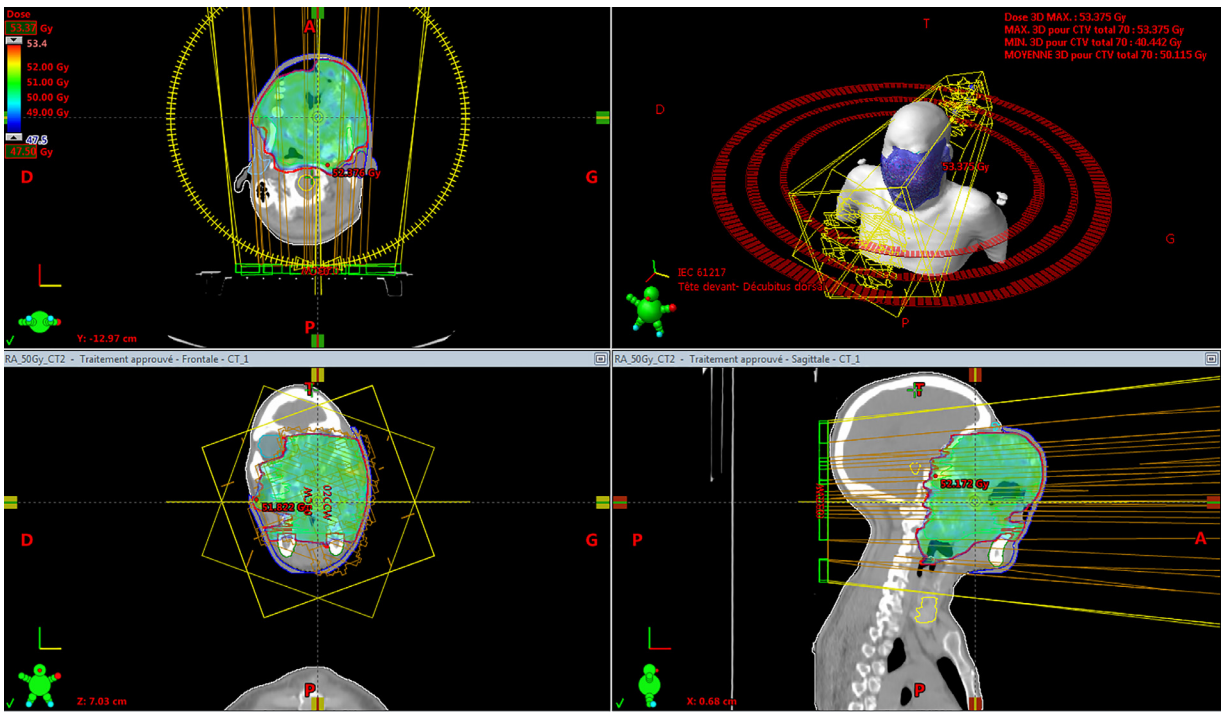

Fig. 5 – Graphical representation of treatment planning using VMAT radiotherapy.

ever, due to the infiltration of the left globe and left optic nerve by the tumor, these structures were excluded from the designation of critical structures in this case.

The facial CT scan conducted 3 months after radiotherapy showed the persistence of the lesion, with a 20% reduction in volume, consistent with stable disease according to RECIST criteria.

A thoraco-abdominal CT scan performed 6 months after the conclusion of treatment revealed the emergence of secondary pulmonary lesions in both lung fields. Based on these findings, the decision was made to initiate palliative chemotherapy. Unfortunately, the patient passed away after the fourth cycle of treatment.

Discussion

Chondrosarcoma is an exceptionally rare malignancy in the head and neck, representing less than 0.1% of tumors in this region. It can affect individuals from 16 months to 74 years, with peak incidence in the third to sixth decades of life [4]. Histogenetically, chondrosarcoma arises from a mature cartilaginous compartment, and its presence in the maxilla, which is predominantly composed of membranous ossification, remains debated [5]. It is theorized that these tumors may originate from the differentiation of primitive mesenchymal cells rather than residual embryonic cartilage.

Clinically, maxillary chondrosarcoma often presents with painless swelling in the maxillary region, gradually leading to facial asymmetry [6]. As the tumor progresses, patients may experience nasal obstruction due to encroachment into the nasal cavity. Initially asymptomatic, localized pain can develop over time, along with dental issues such as loose teeth and discomfort, which may be misinterpreted as dental problems. Involvement of the maxillary sinus can cause sinusitis-like symptoms, including drainage and nasal discharge. If the tumor extends toward the orbit, ocular symptoms like vision changes or proptosis may occur. These manifestations can evolve slowly and often mimic other conditions, contributing to diagnostic delays.

Radiologically, chondrosarcoma exhibits diverse features that aid in diagnosis, though these findings can often be nonspecific [7].

On computed tomography (CT) scans, the lesions typically appear as expansile, lytic areas with variable degrees of calcification. They may be well-defined or exhibit an infiltrative appearance, often extending into adjacent structures like the maxillary sinus. Cortical thinning and destruction of surrounding bone are common observations. Magnetic resonance imaging (MRI) provides detailed insights into the lesion's soft tissue involvement and signal characteristics, generally revealing well-defined masses with heterogeneous signal intensity, reflecting various tumor components, including cartilage and myxoid areas. Conventional radiographs may show chondrosarcomas as radiolucent lesions or demonstrate mixed radiolucent-radiopaque characteristics, with margins that can be well-circumscribed or irregular, complicating differentiation from other lesions. Although bone scintigraphy is not typically used for initial diagnosis, it may indicate increased metabolic activity in the area of the tumor. Due to their heterogeneous nature, chondrosarcomas must be distinguished from other bony lesions such as osteosarcomas, giant cell tumors, and odontogenic lesions, all of which may present with similar radiological features. Collectively, these imaging modalities assist in assessing disease extent, guiding surgical planning, and monitoring treatment response.

Histopathologically, chondrosarcoma is classified into subtypes including conventional, myxoid, and mesenchymal variants [8]. Microscopic examination typically reveals a cartilaginous matrix, often characterized by a lobular architecture [9]. The tumor is composed of malignant chondrocytes displaying varying degrees of cellularity, nuclear pleomorphism, and atypical mitotic figures. In low-grade chondrosarcoma, the matrix appears hyaline and well-differentiated, with relatively few atypical cells. In contrast, high-grade variants show marked cellularity, increased nuclear pleomorphism, and a higher mitotic rate, often with areas of necrosis.

The tumor may also demonstrate myxoid changes, where the matrix contains abundant myxoid stroma, and in some cases, dedifferentiated components can be present. Immunohistochemical analysis often reveals positive staining for S-100 protein, a marker indicative of chondrogenic differentiation, while other markers like keratins and epithelial markers typically show negativity. The differential diagnosis includes conditions such as chondroma, chondroblastic osteosarcoma, and other neoplasms with cartilaginous differentiation.

Chondrosarcoma grading is vital for prognosis and treatment planning, using a 3-tier system [10].

Grade I (low-grade) features well-differentiated cells with minimal nuclear pleomorphism and low mitotic activity, yielding a high 5-year survival rate of about 90%.

Grade II (moderately differentiated) shows increased cellularity and nuclear atypia, with survival rates around 81%.

Grade III (high-grade) presents with significant pleomorphism and high mitotic activity, resulting in a lower 5-year survival rate of approximately 43%. Accurate grading is essential for guiding therapeutic decisions and assessing patient outcomes.

The treatment of chondrosarcoma, especially in the head and neck region, primarily hinges on surgical intervention, which is the cornerstone of management [11]. The main objective of surgical resection is to achieve complete excision of the tumor while preserving surrounding vital structures and maintaining functional integrity. In localized cases, wide local excision is typically performed to secure clear margins, as incomplete resection significantly elevates the risk of local recurrence. In instances where the tumor infiltrates adjacent tissues, more extensive procedures such as partial or total maxillectomy may be required, followed by reconstructive efforts using local or free flaps to restore facial form and function. The role of adjuvant therapies, such as radiotherapy and chemotherapy, remains a topic of ongoing debate. Radiotherapy may be a valuable option in select cases, especially for patients with positive surgical margins or unresectable tumors, as it can help control local disease [12]. However, chondrosarcomas are typically radioresistant, and the close proximity of these tumors to critical structures limits the dose that can be safely administered with conventional radiotherapy [13]. Our

case highlights this radioresistance, as evidenced by the absence of response to the treatment.

Long-term follow-up is essential due to the tendency for local recurrence and late metastasis associated with chondrosarcoma, necessitating regular imaging and clinical evaluations to ensure early detection of any recurrence.

Controlling disease progression in patients with metastatic or unresectable chondrosarcoma remains challenging due to the tumor's resistance to conventional chemotherapy. As a result, there is a pressing need for innovative therapeutic strategies, including targeted therapies and immunotherapy, to enhance clinical outcomes for these patients. Recent research has identified several potential biomarkers and therapeutic targets, such as IDH1/2 and COL2A1, which may guide the development of more effective treatments [14]. A multidisciplinary approach, involving surgical oncologists, radiation oncologists, and pathologists, is vital to optimize treatment strategies and enhance patient outcomes, as prognosis is influenced by tumor grade, size, location, and the completeness of resection.

Conclusion

In conclusion, this case of maxillary chondrosarcoma highlights the aggressive nature of the lesion and the poor prognosis associated with delayed diagnosis. The significant clinical symptoms and imaging findings underscore the necessity for timely evaluation of facial swellings, as early intervention is crucial in improving outcomes. Awareness of the potential malignancy in such cases is essential to prevent diagnostic delays and ensure appropriate treatment.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

REFERENCES

[1] Ngo QX, Ngo DQ, Tran TD, Le DT, Van Le Q. Chondrosarcoma of the Maxilla. Ear Nose Throat J 2022;101(1):NP6–7. doi:10.1177/0145561320942358.

[2] Brimioulle M, Bowles PF, Pelser A. Maxillary chondrosarcoma mimicking torus palatinus. BMJ Case Rep 2017;2017:bcr2017221629. doi:10.1136/bcr-2017-221629.

[3] Shah KD, Ezzy MK, Patekar SK, Bradoo R. Chondrosarcoma of the maxilla. BMJ Case Rep 2023;16(11):e253143. doi:10.1136/bcr-2022-253143.

[4] 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(2):139–43. doi:10.1016/0030-4220(91)90454-k.

[5] Ram H, Mohammad S, Singh G, Singh SV. Chondrosarcoma of body of the mandible. Natl J Maxillofac Surg 2013;4(2):242–4. doi:10.4103/0975-5950.127661.

[6] Cuevas-González JC, Reyes-Escalera JO, González JL, Sánchez-Romero C, Espinosa-Cristóbal LF, Reyes-López SY, et al. Primary maxillary chondrosarcoma: a case report. World J Clin Cases 2020;8(1):126–32. doi:10.12998/wjcc.v8.i1.126.

[7] Jang BG, Huh KH, Kang JH, Kim JE, Yi WJ, Heo MS, et al. Imaging features of chondrosarcoma of the temporomandibular joint: report of nine cases and literature review. Clin Radiol 2020;75(11):878.e1–878.e12. doi:10.1016/j.crad.2020.07.016.

[8] Gazendam A, Popovic S, Parasu N, Ghert M. Chondrosarcoma: a clinical review. J Clin Med 2023;12(7):2506. doi:10.3390/jcm12072506.

[9] Ismail A, Boujguenna I, Hattab K, Mansouri N, El Ganouni NCI, Idrissi MO, et al. A cartilage-forming tumor of the mandibular angle: a case report. J Med Case Rep 2022;16:176. doi:10.1186/s13256-022-03359-x.

[10] Sbaraglia M, Bellan E, Dei Tos AP. The 2020 WHO classification of soft tissue tumours: news and perspectives. Pathologica 2021;113(2):70–84. doi:10.32074/1591-951X-213.

[11] Mahajan AM, Ganvir S, Hazarey V, Mahajan MC. Chondrosarcoma of the maxilla: a case report and review of literature. J Oral Maxillofac Pathol 2013;17(2):269–73. doi:10.4103/0973-029X.119759.

[12] Lyoubi M, Oukessou Y, El Krimi Z, Roubal M, Mahtar M, Reguragui M, et al. Aggressive Mesenchymal chondrosarcoma of the maxilla: case report. Int J Surg Case Rep 2022;91:106696. doi:10.1016/j.ijscr.2021.106696.

[13] Gilbert A, Tudor M, Montanari J, Commenchail K, Savu DI, Lesueur P, et al. Chondrosarcoma resistance to radiation therapy: origins and potential therapeutic solutions. Cancers (Basel) 2023;15(7):1962. doi:10.3390/cancers15071962.

[14] Miwa S, Yamamoto N, Hayashi K, Takeuchi A, Igarashi K, Tsuchiya H. Therapeutic targets and emerging treatments in advanced chondrosarcoma. Int J Mol Sci 2022;23(3):1096. doi:10.3390/ijms23031096.