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Case report

Aggressive Mesenchymal chondrosarcoma of the maxilla: Case report

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ABSTRACT

Introduction: Chondrosarcomas of the head and neck are extremely rare, accounting for approximatively 0.1% of all head and neck malignant tumors. Mesenchymal chondrosarcomas are particularly aggressive, with a high propensity for recurrence.
Case report: We report the case of a 25 years old man with no prior medical history, who presented to our facility with an aggressive tumor of the left maxilla. Upon radiologic and histopathological examinations, the tumor was revealed to be a mesenchymal chondrosarcoma. The patient was treated using surgical excision of the tumor, neoadjuvant chemotherapy and adjuvant radiotherapy.
Conclusion: Due to the high risk of recurrence and the possibility of metastasis occurring long after the initial diagnosis, it is important that patients with mesenchymal chondrosarcomas receive close and regular follow-ups after treatment.

1. Introduction

Chondrosarcoma is a malignant tumor derived from a cartilaginous origin, that tends to maintain its cartilaginous nature through its evolution [1]. This neoplasm occurs mostly in the long bones [2]. Chondrosarcomas arising in the head and neck region are considered rare, and account for 0,1% of all head and neck tumors, and for 1 to 12% of all chondrosarcomas [2–5].

The most frequent sites of chondrosarcomas in the head and neck are the maxilla, the mandible and the sino-nasal region [6,7]. In this article, we report the case of a mesenchymal chondrosarcoma of the maxilla in a 25 years old patient. This case has been reported in line with the SCARE 2020 criteria [8].

2. Case report

A 25 years old man, with no significant medical history and no history of alcohol or drug abuse, presented at our department with a 6-month history of bilateral nasal obstruction, epistaxis and anosmia. These symptoms were initially located only in the left nasal fossa, but became bilateral and persistent as they progressed. The evolution was marked a few weeks prior to the consultation by the sudden onset of visual loss, diplopia, proptosis and pain, all in the left eye, associated with a swelling of the face and the palate. The patient also reported bilateral hearing loss, as well as a severe deterioration of his general well-being, with a weight loss of 13 kg since the beginning of the symptoms.

Clinical examination revealed a large ulcerated tumor in the left nasal fossa, causing a prominent bulge of the entire palate (Fig. 1). At this stage, the patient's quality-of-life and general well-being was already severely altered, with a performance score deemed at stage 2.

Magnetic resonance imaging (MRI) and computed tomography (CT) were performed to assess the nature and extension of the nasal mass. The CT scan showed the presence of a tumoral process developing at the expense of the left maxilla, filling the maxillary sinus and responsible for the destruction of its anterior and medial walls (Fig. 2). This tumor also invaded the ethmoïdo-nasal region, the sphenoid sinus, the naso-pharynx, and part of the oropharynx (Fig. 3). It measured 60x55x84 mm, and contained calcifications within it. The upper end of this process extended to the inferior wall of the orbit, with no evidence of intra-orbital extension or intracranial extension.

The MRI scan found that this mass had a low signal intensity on T1 weighed images, a high signal intensity on T2 weighed images, and was enhanced in a heterogeneous on the sequences taken after administering gadolinium (Fig. 4).

A biopsy of the mass was performed under local anesthesia.

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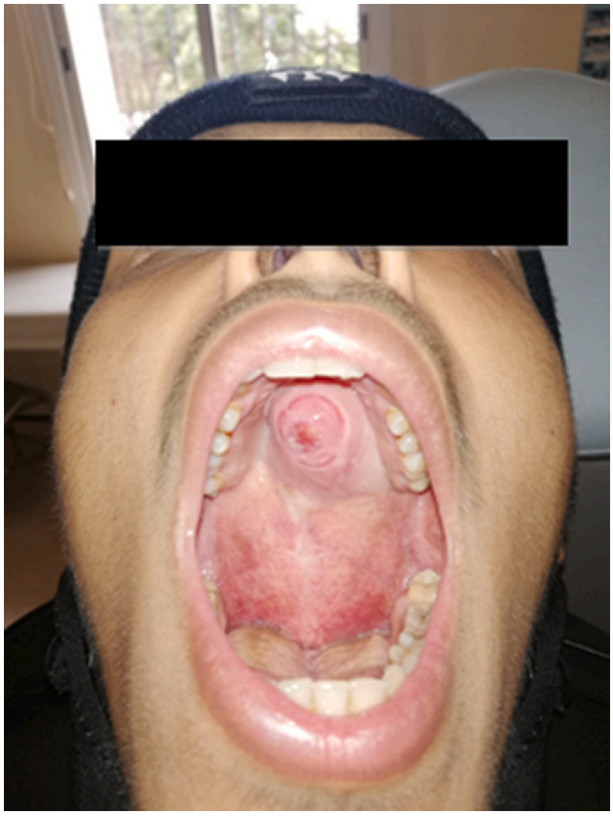

Fig. 1. Image of the tumor bulging in the entire palate.

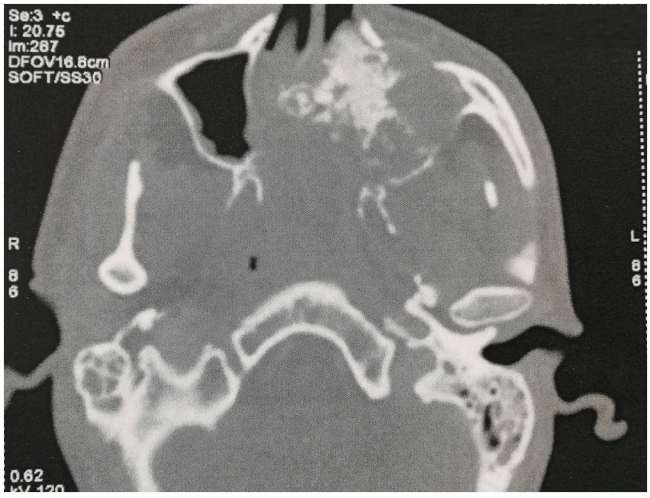

Fig. 2. Axial CT image showing a tumoral process filling the left maxillary sinus and destroying its medial and anterior walls.

Pathological study of the samples showed histologic findings consistent with mesenchymal chondrosarcoma.

The case of this patient was discussed in a multidisciplinary meeting between ENT specialists and oncologists, the decision was for the patient to receive neoadjuvant chemotherapy first, to reduce the tumor volume and allow surgical excision.

After receiving 4 chemotherapy treatments, the patient underwent surgery, which consisted of a left maxillectomy and tumor resection performed by a seignor surgeon with several decades of experience. Since the tumor was sizeable, the resection was made in several fragments, with the largest piece measuring 8.5x6x3 cm. Histopathological

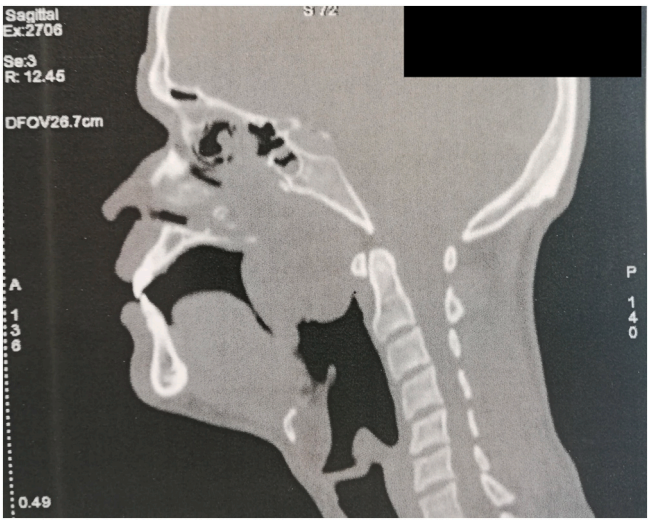

Fig. 3. Sagittal CT image showing the extension of the tumor to the ethmoid, the nasopharynx and the oropharynx.

examination of the resection material confirmed the diagnosis of an intermediate grade chondrosarcoma, and the margins of excision were timorous (Fig. 5). The postoperative consequences were simple with oral feeding through a nasogastric tube. Psychological support was necessary for the acceptance of the disfigured face of the patient the time to rehabilitate. The patient was addressed to a radiotherapy facility, where he received subsequent radiation therapy.

The first post-operative check-up took place 15 days after surgery. The patient had not begun radiation therapy yet, and the maxillectomy cavity was in good condition. The 3-month follow-up found the patient in a fairly good condition, with no sign of local recurrence. The follow-up examination of our patient will be long-term, a close rhythm the first 5 years at the rate of a consultation every 3 months the first 2 years, then every 6 months up to 5 years then once a year. It will be based on clinical examination, nasal endoscopy and CT imaging. The detection of a recurrence will require a discussion in a multidisciplinary meeting.

The patient had difficulty accepting the surgery since it is a heavy gesture with several consequences on his quality of life.

3. Discussion

Maxillofacial chondrosarcomas are extremely rare [1]; they account for less than 2% of all head and neck tumors, and represent approximatively 10% of all chondrosarcomas in literature [9–11]. Head and neck chondrosarcoma can grow from any bone, cartilaginous or soft-tissue structure, but it most commonly affects the maxilla and the mandible [9,10,12].

The epidemiologic characteristics of these tumors are not well known, in part due to their rare occurrence. The male-female sex ration is roughly 1.2:1 [12]. Chondrosarcomas occur most commonly in patients younger than 40 years old [9,13]. This matches with our case, where the patient was a 25 years old man. An association with medical conditions, such as leukemia, melanoma, Paget disease or fibrous dysplasia, has been described in several instances [9–12]. However, this was not the case with our patient, who has no known medical history.

Some studies have suggested that the pathogenesis of chondrosarcoma is related to the inactivation of known tumor suppressor genes, such as p16 and p53 genes [9,14–16].

Maxillary chondrosarcoma generally presents as a painless tissue mass in the nasal cavities. Other symptoms may be associated, depending on the degree of extension of the tumor, such as nasal obstruction, anosmia, epistaxis, impaired vision, swelling of the cheek or of the palate, or dental abnormalities [5,9,12,17]. In our case, the tumor

was discovered at a very advanced stage; that is why the patient presented with all these symptoms on his first consultation.

Radiologic imaging may be highly suggestive of the diagnosis. Typically, chondrosarcoma appears on CT scan as a lobulated mass containing a chondroid matrix, and responsible for the destruction of the adjacent bone structures. The density of the chondroid matrix is lower than the bone matrix; however, some zones of bone density may be found because of localized ossification [9]. MR imaging, the chondroid matrix has a high signal intensity on T2 weighted images, while the bone matrix has a low signal intensity [5,18]. On T1 weighted images, after administering contrast material, the enhancement of the tumor is shown to be heterogeneous: the fibrovascular tissues are enhanced, whereas the cartilaginous tissues, the mucoid tissues and the necrosis are not [19]. This is consistent with the findings in the CT scan and MRI performed on our patient.

Definite diagnosis of mesenchymal chondrosarcoma is established by histopathological examination of biopsy fragments. Microscopic examination typically shows a proliferation of hyaline cartilage in nodules or islands, against a background of packed spindle-shaped or round cells [1,5]. Immunohistochemistry helps confirm the diagnosis, by highlighting the expression of S-100 protein in the cartilage, and CD99 in the round cells [20,21].

The choice treatment for chondrosarcomas of the head and neck is usually radical surgical excision [1,22,23]. Radiation and chemotherapy can eventually be used in addition to surgery, but have been reported to be ineffective [24]. Radiotherapy is generally used particularly when the risk of recurrence is high, when secure margins were not obtained, or when the tumor is judged beyond surgical treatment [5,25–28]. Because of the voluminous size of the tumor, our patient received neoadjuvant chemotherapy, followed by surgical resection and adjuvant radiation therapy.

The prognosis of chondrosarcomas seems to be related to the location of the tumor. Because of its complex location, which often does not allow for a complete excision, and its known aggressive local behavior, maxillary chondrosarcoma is believed to have a high rate of recurrence [1,29]. Other important prognostic factors of long-term evolution include the quality of the surgical resection, and the histological grade of the neoplasm (low, intermediate or high grade). The prognosis is considered good for low and intermediate grade chondrosarcoma [30]. Radiotherapy is reported to improve the prognosis in patients with tumoral margins after surgery [25,31,32]. The mesenchymal subtype of chondrosarcoma is considered more aggressive, and subsequently has a poor prognosis compared to other histological subtypes [5,10,31,33].

In this case, the patient had intermediate grade mesenchymal chondrosarcoma, and the surgical resection was not complete, so his prognosis was not good. Since the risk of recurrence is quite high and metastasis can occur many years after the initial diagnosis, the patient will continue to undergo close and regular check-ups.

4. Conclusion

Head and neck chondrosarcomas are rare malignant tumors, arising most frequently from the maxilla and the mandible. Diagnosis is based on histopathological examination of biopsy material, and is often aided by CT imaging and MRI. Complete surgical resection of the tumor is the only truly effective treatment, although chemotherapy and radiotherapy can sometimes be used to enhance the efficiency of surgery. Because of its poor prognosis and the risk of recurrence and metastasis, long-term follow-up is required for patients treated for mesenchymal chondrosarcoma of the maxilla.

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Ethical approval

Ethical approval has been exempted by my institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Registration of research studies

N/A.

Guarantor

Lyoubi Mouna.

CRediT authorship contribution statement

All the authors worked in coordination to ensure the best management of the disease from diagnosis to surgery with close postoperative monitoring. The authors wrote this article together.

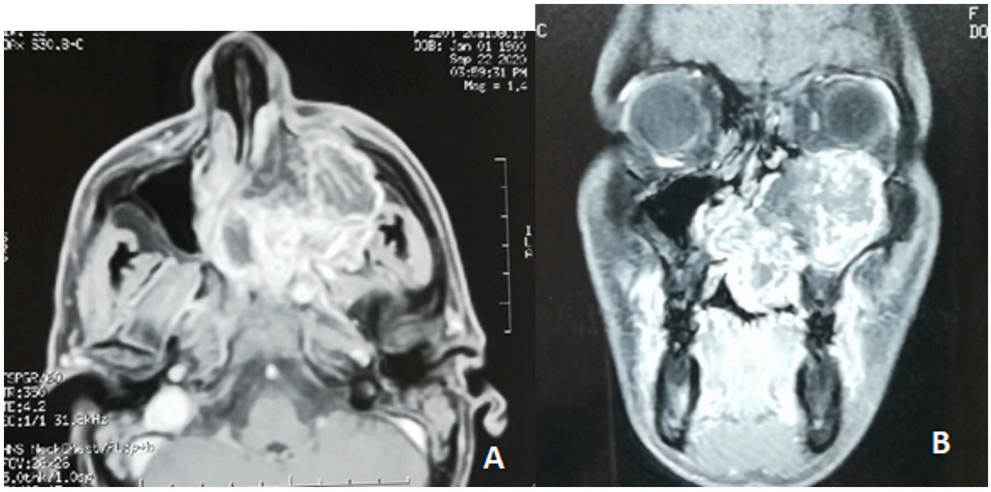

Fig. 4. Axial (A) and coronal (B) T2 weighted MRI images showing the high signal intensity of the maxillary tumor.

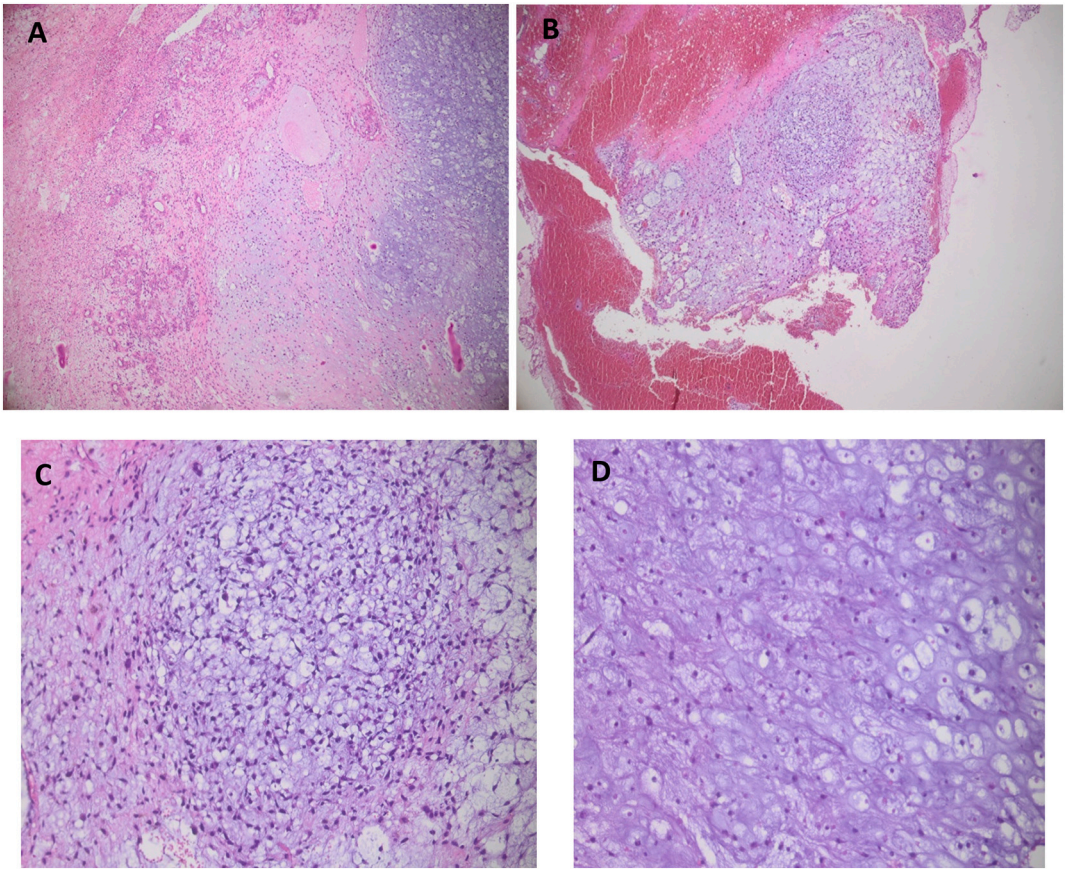

Fig. 5. Images of the histological aspect of the tumor. A: HE x 4, Conventional Chondrosarcoma; B: HE x 4, Tumor proliferation takes on a myxoid appearance with the presence of stellate cells with dense nuclei within a myxoid material; C: HE x 20, Tumor cells exhibit large nuclei, densified with images of bi and multi-nucleation, Mitotic figures are numerous with abnormal mitoses; D: HE x 20, Tumor cells exhibit large nuclei, densified with images of bi and multi-nucleation. Mitotic figures are numerous with abnormal mitoses.

Declaration of competing interest

No conflicts of interest.

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