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CASE REPORT

Chondrosarcoma of the Nasal Cavity and Paranasal Sinuses: A Rare Entity

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ABSTRACT

Chondrosarcomas, slow-growing malignant tumors originating in cartilaginous structures, usual manifest in the head and neck region, with a preference for the maxillofacial skeleton, particularly the mandible and maxilla. However, chondrosarcoma of the sinonasal tract is exceptionally rare, and there are only few case reports.

We detail the case of a 43-year-old woman who incidentally discovered chondrosarcoma in the maxillary and ethmoid sinuses, with nasal extension. Subsequently, the patient underwent surgery via a para-lateral-nasal approach with adjuvant radiotherapy.

The purpose of this study was to describe the clinical findings, management and outcome of sinonasal tract chondrosarcoma.

The primary treatment modality for chondrosarcomas remains surgery, with a transnasal endoscopic approach that offers a viable option for complete resection in select cases.

KEYWORDS

Chondrosarcoma; Nasal cavity; Paranasal sinuses; Treatment

INTRODUCTION

Chondrosarcomas (CSs) rank as the third most-common primary malignancy of bone after myeloma and osteosarcoma. Although the histological appearances of this tumor are characteristics, a differential diagnosis of chondroma, chordoma, and chondromyxoid fibroma should be considered.

Rarely, it can develop in the craniofacial bones, accounting for 8% of all head and neck sarcomas and only 0.1% of all head and neck malignancies with a higher incidence among women and individuals under 50 years of age [1]. In the head and neck, CSs commonly manifest in the mandible, paranasal sinuses, maxilla, and occasionally nasal septum. The diagnosis typically involves imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI), complemented by biopsies to confirm the malignant nature of the tumor [2]. Occurrence of chondrosarcoma in the nasal cavity is uncommon, with less than 200 cases reported in the literature dating from 1950.

Sinonasal tumors are heterogeneous group of tumors arising from different tissues in the area of nasal cavity, paranasal sinuses, and skull base. Diagnosis of cartilaginous tumors is challenging considering varying histopathology and clinical behavior and is based on histopathological analysis.

Chondrosarcomas of the paranasal sinuses pose a compelling diagnostic and treatment challenge. Very rarely will they initially present with ocular symptoms as with our patient. A comprehensive range of medical investigation enables timely recognition and treatment.

The purpose of this study was to describe the clinical findings, management and outcome of sinonasal tract chondrosarcoma.

CASE REPORT

A 43-year-old woman presented a history of chronic left lacrimation lasting 4 months. It was painless and there was no history of blurred vision, prolonged fever, neck mass, or any associated symptoms of the ear or throat. The ophthalmological examination revealed a nonreducible axial left exophthalmos (Figure 1) that was nonpainful, with impermeability of the nasolacrimal duct, without diplopia or decrease visual acuity. Nasal endoscopy showed a tumor process in the left nostril that originated from the middle-left meatus, with a nonbleeding pinkish white appearance, destroying the nasal septum, and extending to the right nostril. It appeared to have a smooth surface with prominent vessels. Neck examination did not reveal palpable lymph nodes. Neurological examination, particularly of the cranial nerves was normal.

Figure 1: A non-reducible axial left exophthalmos.

An orbital MRI was performed, revealing the presence of a heterogeneous expansive process (68 mm × 61 mm × 57 mm) with irregular contours. The process included the ethmoidal cells and the left maxillary sinus, prolapsed into the nasolabial and oropharyngeal lumen by filling the choanae and the ipsilateral nasal fossa. It exhibited heterogeneous hyposignal T1, hypersignal T2 (Figure 2), with weak signal areas in all sequences. It was heterogeneously enhanced after the injection of gadolinium. The MRI showed significant locoregional extension with invasion of the left infratemporal fossa and parapharyngeal space. It came into contact with the floor of the sphenoid sinus with contrast enhancement. It destroyed the nasal septum in its posterior part, the sieve blade, and the posterior part of the hard palate, and infiltrated the soft palate. There was the presence of lysis of the left papyraceous lamina and destruction of the inferior and medial walls of the left eye, with the left endo-orbital extension responsible for grade I exophthalmos. The optic nerve was displaced laterally with the loss of the fatty safety line. No detectable signs of endocranial invasion were observed.

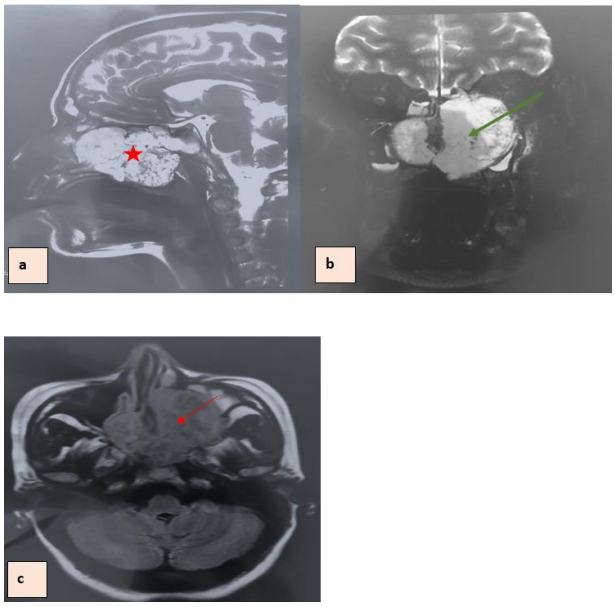

Figure 2: An orbital MRI in sagittal **(a)** and axial **(b, c)** section: Showing a heterogeneous expansive process with irregular contours. The process invaded the ethmoidal cells, the left maxillary sinus, infratemporal fossa and parapharyngeal space (red color arrow), prolapsed into the nasolabial and oropharyngeal lumen. It exhibited heterogeneous hyposignal T1(c), hypersignal T2 (a, b). The MRI showed invasion of the It destroyed the nasal septum in its posterior part, the sieve blade, the posterior part of the hard palate (red color star) and left endorbital extension (green color arrow).

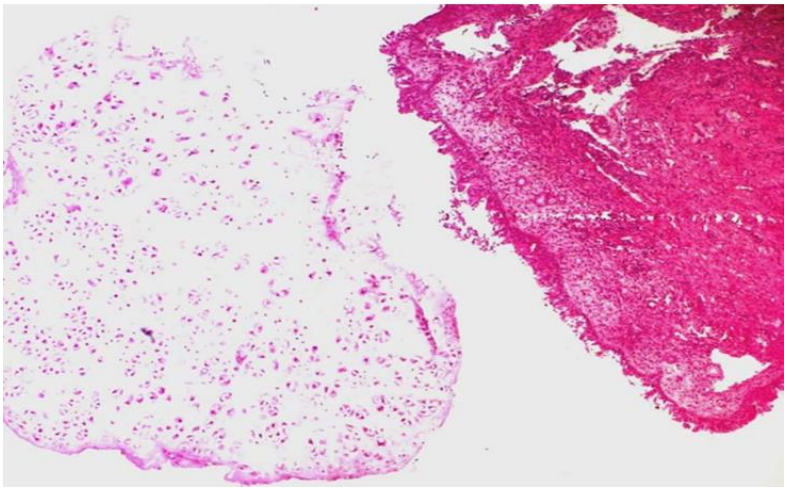

Figure 3: One fragment involved a malignant tumor of cartilaginous differentiation with lobulated growth and slightly increased cellularity, and the second fragment involved a nasal mucosa with acute inflammatory remnants (Hex50).

A biopsy was performed under local anesthesia, and histological and histochemical studies concluded that it was a well-differentiated chondrosarcoma (CS) (grade 1) with malignant chondrocytes show small, round hyperchromatic nuclei and occasional binucleated cells, mitoses are absent (Figure 3 - Figure 5).

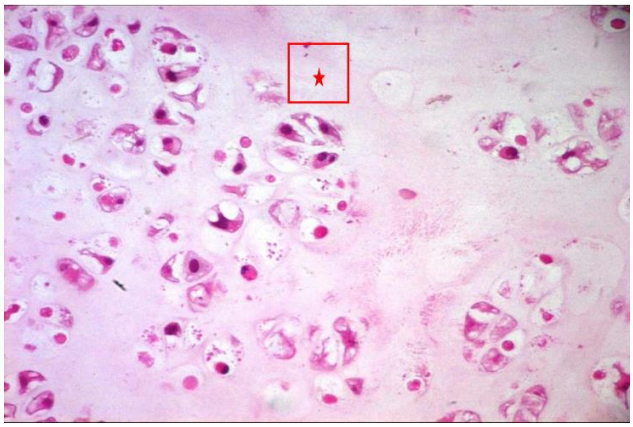

Figure 4: Malignant chondrocytes show small, round hyperchromatic nuclei and occasional binucleated cells (red color star), mitoses are absent (Hex200).

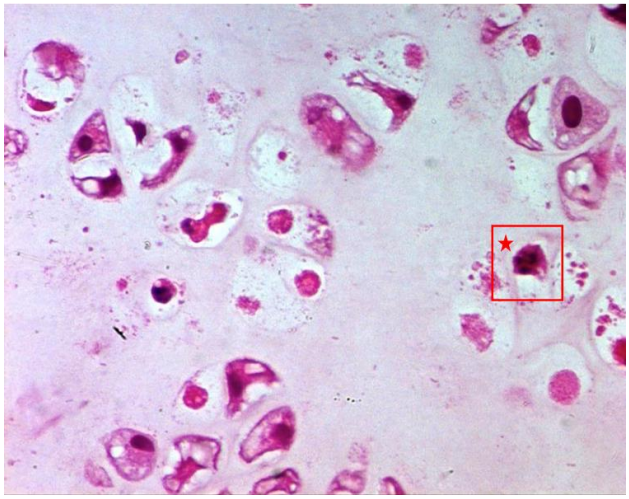

Figure 5: Malignant chondrocytes show small, round hyperchromatic nuclei and rare mito-necrosis (red color star) (Hex400).

The case was discussed in the tumor board and it was decided to undergone surgery in the form of paralateral-nasal approach followed by adjuvant therapy. The patient underwent surgery via a para-lateral-nasal approach, with incomplete resection due to endo-orbital extension. Weber Ferguson incision was performed. The tumor was found to fill both nasal cavities with involvement of the left maxillary sinus and left ethmoid cells. It caused lysis of the left lamina papyracea and extending into the left orbit without invasion of the orbital adnexa. Tumor was infiltrating the hard palate on the left side. The lesion extended to the nasal septum. Bilateral medial maxillectomy was done. There was no defect of hard palate after excision of the lesion.

The excised specimen was sent to the pathologist, confirming the diagnosis. Postoperative recovery was uneventful. The patient received adjuvant radiotherapy. The patient was then lost to view.

DISCUSSION

Chondrosarcoma is characterized by its malignant nature and the production of cartilage matrix. It represents 20% to 27% of all primary bone sarcomas at various anatomical locations [2]. Typically found in long bones and pelvic

bones, CS occurs in 5% to 10% of cases in the head and neck region [3]. This slow-growing tumor exhibits a preference for males, with the highest frequency occurring between the ages of 40 and 60 [4]. Our patient was a 43-years-old woman.

The occurrence of CS in the paranasal sinus system is exceptionally rare. Notably, Knott et al.'s series [5], spanning 25 years, describes 13 cases at this location. Other studies, such as Touati et al. (2 cases) [4], Krömer et al. [3], and Rowley et al. [6] (1 case each), also report limited instances. The clinical presentations of CS depend on the extent of the tumor. In the sinonasal region, nasal obstruction is the predominant symptom, accompanied by headaches, epistaxis, anosmia, facial pain, signs of cranial nerve involvement or visual disturbances [7], like our patient how consulted for chronic left lacrimation and nonreducible axial left exophthalmos.

Imaging techniques, primarily CT and MRI, play a crucial role. CT provides detailed information on bone destruction, which includes structures such as the ethmoid blade, orbit walls, and the bony palate. Sinonasal CSs tend to reshape the sinonasal cavity, with the degree of bone erosion correlating with tumor grade. Well-differentiated tumors exhibit punctate cartilaginous calcifications.

On MRI, the cartilaginous matrix of CSs displays very low signal intensities on T1- and T2-weighted images, with homogeneous or heterogeneous enhancement post-contrast injection. Calcified and ossified regions exhibit very weak signals across all sequences [8,9]. MRI helps detail the extension into surrounding soft tissues and distinguishes between granulomatous tissue and recurrences during postoperative monitoring of CS.

Diagnosis of the tumor is made on the basis of histopathological features. The most prevalent form is Conventional CS, which can be categorized into four degrees of malignancy based on cell abundance, nuclear polymorphism, and mitotic rate. Other forms include clear cell CS, mesenchymal CS, and undifferentiated CS [3]. In our case the chondrosarcoma was categorized as grade 1. Cause for chondrosarcoma is still unknown despite proposed etiological factors such as trauma, previous exposure to irradiation, asbestos, beryllium, Teflon, aluminium, iron, or radioactive isotopes. Twelve to thirty-eight percent of chondrosarcomas develop secondary to previous conditions such as solitary or multiple exostosis, Maffucci’s syndrome, Paget’s disease, Ollier’s disease, and fibrous dysplasia, most are sporadic lesions without any known causative factor [3].

Surgical excision in bloc resection is the primary treatment approach because it is relatively radioresistant [10,11]. The surgical approach depends upon location, extension, and histological grade of the tumor. Obtaining soft surgical margins is the key point for local control [9].

Adjuvant treatment often involves radiation, rarely used in isolation. The recommended dose for radiation therapy is 6000 cGy to 7000 cGy, administrated in 30 to 35 fractions, while chemotherapy is specifically reserved for high-grade cases [12]. In our case the patient received, surgical excision as a primary treatment followed by an adjuvant radiotherapy. Postoperative radiation therapy is used in instances of sizable tumors, involvement of skull base or vital neurovascular structure, and subtotal resection, as seen in our patient. Chondrosarcoma survival rates range from 44% to 87%, with respectability identified as a crucial prognostic factor, since most deaths are the result of locally invasive disease [1]. A systematic review indicates that patients undergoing radiation therapy exhibit a lower incidence of local recurrence and distant metastases compared to those treated with surgery alone

(29.4% *vs.* 32.8%). Surgery is the treatment of choice. Given the low incidence of head and neck cancers and the complex anatomy of the head and neck region, management should be ideally centralized in specialized centers with dedicated multidisciplinary teams.

However, the limited sample size due to the rarity of the lesion and the absence of standardized patient data pose challenges in drawing definitive conclusions [10].

CONCLUSION

Chondrosarcomas that occur in the sinonasal tract are uncommon. Patients who receive an early diagnosis and undergo appropriate surgical intervention tend to have a more positive prognosis. The primary cause of mortality is unmanageable local disease that led to the compression of neighbouring vital structures.

AUTHOR CONTRIBUTIONS

Malika EL Omri and Maroua Ben Njima drafted the manuscript. All authors read and approved the final version of the manuscript.

DATA AVAILABILITY STATEMENT

Not applicable

CONFLICT OF INTEREST

The author declares that the research was conducted without any commercial or financial relationships that could be construed as a potential conflict of interest.

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INFORMED CONSENT STATEMENT

The patient gave their informed consent for inclusion before they participated in this study.

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