



Diagnostic and therapeutic difficulties in sphenoid chondrosarcoma: A case reports


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Abstract

Chondrosarcoma is a rare tumor, and sphenoidal localization is exceptional.

Our work highlights the difficulties of diagnosis, given the non-specificity of the clinical and radiological picture, hence the importance of anatomopathological examination to establish the diagnosis and classify the patient for a good prognostic evaluation.

In this article, we report the case of a 54-year-old diabetic woman on oral antidiabetics and hypertensive on thiazides who presented with bilateral nasal obstruction, more marked on the left, with headaches and anterior rhinorrhea. Nasal endoscopy revealed the presence of a process in the left nasal fossa.

CT and MRI confirmed the presence of a malignant-looking mass that required excision, which was not complete given the difficult localization. This allowed tumor reduction and diagnosis thanks to anatomopathological examination, which remains the key examination in this case.

Chondrosarcoma of the sphenoid is rare and difficult to diagnose, manage and treat, as the picture is non-specific and the location inaccessible to surgery, and the prognosis depends on the stage and quality of excision.

Key words: chondrosarcoma, sphenoid, rhinorrhea, endonasal surgery, case report

Introduction

Chondrosarcoma is a primary malignant tumor of bone that produces tumor cartilage without ever developing tumor bone tissue. [1].

The tumor may arise de novo (primitive chondrosarcoma) or develop on a pre-existing lesion (10% of cases). [2].

Progression is marked by local recurrence and metastases, particularly in the lungs, sometimes occurring very late [3]. [4]. [5].

Clinical case

Patient information: 54-year-old woman, diabetic on ADO, hypertension on thiazide, hospitalized for total left nasal obstruction associated with right nasal obstruction, clear anterior rhinorrhea homolateral to nasal obstruction, nocturnal mouth breathing, and headache, without dysosmia or epistaxis with abolition of left-sided nasal flow.

The Nasal endoscopy showed: serous secretions in the left nasal cavity, deviation of the nasal septum to the right, bluish polypoid mass budding, pedunculated in the sphenoidal sinus filling the left nasal cavity, collapsing into the cavum and partially obstructing the right nasal cavity.

A naso-sinus CT scan showed an iso-dense process in the left choanal orifice extending into the sphenoid sinus, relative obstruction of the right choanal orifice with destruction of the vomer implantation base and thickening of the upper wall of the nasopharynx (Figure 1).

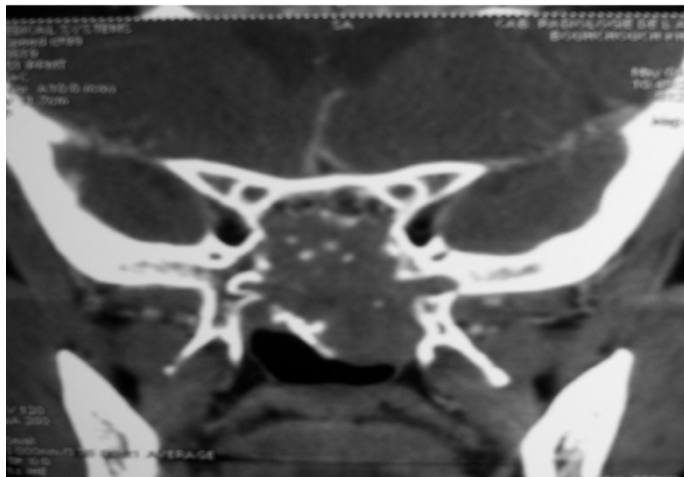


Figure 1:Nasosinus CT scan showing a heterogeneous process with calcifications on the sphenoid obstructing the cavum

An MRI scan showed a heterogeneous, lobulated, expansive lesion process, enclosing partitions with calcifications occupying the sphenoid body and extending towards the 2 choanae, with filling anteriorly and superiorly of the posterior part of the ethmoidal cells. Below, lysis of the posterior part of the nasal septum and tumour pushing back the inferior turbinates (especially on the left), the 2 carotid arteries are pushed back but permeable. (Figure 2)



Figure 2 :Nasosinus MRI showing a heterogeneous, lobulated, expansive lesion process, containing partitions with calcifications occupying the sphenoid body and extending towards the 2 choanae.

The Treatment consisted of endonasal excision of fragments to unblock the choana and left nasal cavity. Histological findings were consistent with grade 2 chondrosarcoma, and the patient was referred for radiotherapy. A thoraco-abdomino-pelvic CT scan for extension was negative.

The patient reported a marked improvement in clinical signs such as nasal obstruction and rhinorrhea, which disappeared completely, along with headaches.

The anatomopathological study was consistent with a chondrosarcoma of the sphenoid, and the patient was referred to complete her treatment with radiotherapy. The patient was lost to follow-up despite our attempts to contact her. We never heard back from her.

Discussion

Chondrosarcoma is a malignant tumor, with head and neck localization accounting for 12%. Sphenoidal localization is extremely rare.

Its incidence increases with age, peaking between the ages of 40 and 60, with a clear predominance of females (3 females to 2 males).

Symptoms are varied, depending on the extent of the tumor. There may be ocular signs such as reduced visual acuity, retro-ocular pain, visual fog, headaches, endocrine signs due to compression of the pituitary stem, and rhinological signs such as nasal obstruction and rhinorrhea[6].

Medical imaging provides characteristic, albeit unspecific, signs. This patient's CT scan showed an isodense process in the left choanal orifice extending into the sphenoidal sinus, and relative obstruction of the right choanal orifice.

Magnetic resonance imaging (MRI) is the key examination in the management of chondrosarcoma, enabling us to study the tumor's relationship to surrounding structures, to make a differential diagnosis with other tumors, and to determine the lobulated nature, presence of calcification and peripheral enhancement, all of which are signs in favor of chondrosarcoma. MRI angiography may be useful for assessing relationships with vessels.

Anatomopathological examination is not straightforward, as it poses a problem of differential diagnosis with chondroma. In this context, Evans has distinguished three grades of increasing aggressiveness [7].

Grade I

Boundaries with chondromas are imprecise, which explains the diagnostic difficulties.

The chondroid substance is well differentiated.

Cell density is low, except at the periphery of the lobules.

Grade II

Cell density is higher, with binucleated cells representing a maximum of 10% of the cell population.

Grade III

Clear cytonuclear atypia with poor, inhomogeneous, poorly calcified chondroid substance.

Chondrosarcoma is treated surgically, but complete removal of the tumour is far from easy, due to the extent of the disease. Chondrosarcoma is not radiosensitive, and chemotherapy has shown no efficacy. [8]

However, some authors recommend complementary radiotherapy at a dose of 50-60 Gy after the third course of chemotherapy.

The prognosis remains poor, depending on the type, histological grade, extension, location and quality of excision.

Survival rates vary from 44% to 81% for chondrosarcomas of the head and neck region (9). The evolution is marked by local recurrence and distant metastases, which are mainly pulmonary and occur very late [3], [4], [7], [8], [9].

The 10-year survival rate is less than 30% for all bony and extra-skeletal localizations.

Conclusion:

Chondrosarcoma = primary malignant tumor of bone producing tumor cartilage [1], can arise de novo (primary chondrosarcoma) or develop on a pre-existing lesion (10% of cases).

Sphenoidal localization is relatively rare.

Poor prognosis depending on grade

Patient perspective:

Before admission and in the operating room, the patient received an explanation of the procedure in order to obtain her consent. Objective abnormalities and risks associated with anesthesia and surgery were explained.

Informed consent :

A written informed consent, dated and signed, was obtained from the patient.

Conflict of interest

The authors declare no conflict of interest.

Authors' contributions

All authors have contributed to the management of the patient and have read and approved the final version of the manuscript.

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