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# Chondrosarcoma in petroclival synchondrosis without visual change. A case report

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## ABSTRACT

Chondrosarcomas are a heterogeneous group of malignant bone tumours that share the production of the chondroid matrix in common. At the base of the skull, they are most commonly found in the region of the various synchondroses with an affinity for the petroclival fissure, they are locally invasive tumours, with little capacity to perform metastasis. The age group affected is variable, however, they frequently occur in middle-aged adults. Its clinical manifestation depends on the location and local extent; headache or paralysis of cranial nerves, particularly of the VI nerve is a frequent sign. As the petrous apex cannot be viewed directly, imaging studies such as computed tomography and magnetic resonance imaging play an important role in the evaluation of injuries. We present a case of a 36-year-old patient with chondrosarcoma of petroclival syndromes without visual changes. For the identification of this pathology, a battery of imaging tests was used and the diagnosis was made assertively, preserving the best choices for the treatment of the patient.

## INTRODUCTION

Chondrosarcomas (CS) is a heterogeneous group of malignant bone tumors that share in common the production of the chondroid (cartilaginous) matrix. Cartilaginous tumors are almost always found in bones that arise from endochondral ossification, noting in the growth plate, the proliferation and hypertrophic cell differentiation of chondrocytes, which undergo apoptosis, followed by subsequent local invasions by vessels and osteoblasts, thus initiating the matrix formation, which consequently leads to longitudinal bone growth. Chondrosarcomas of the skull are rare and have a higher incidence in

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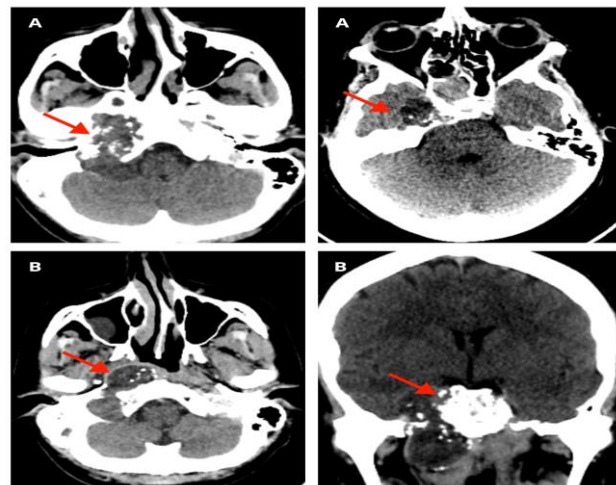
male patients in a 2:1 ratio. They are usually diagnosed around the third and fourth decades of life(1,2).

At the base of the skull, they are most commonly found in the region of the various synchondroses that remain after ossification of the embryonic chondroid matrix, with an affinity for the petrooccipital (petroclival) fissure. Therefore, they present as para-sagittal tumors. These tumors are locally invasive, with little capacity to metastasize. They spread through the petroclival fissure, involve the clivus, the petrous portion of the temporal bone, and are more prominent in cisterns or soft tissues at the base of the skull (5,6).

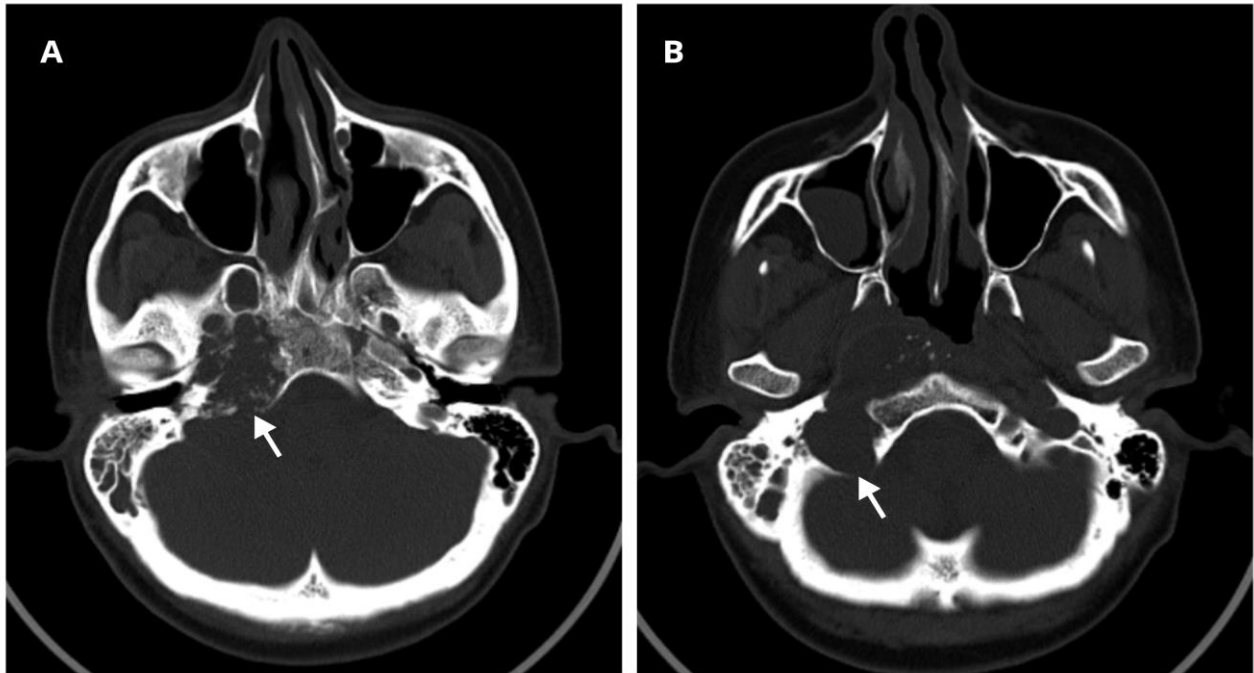
### CASE REPORT

A 36-year-old female patient enters a private clinic in the city of Dourados-MS-Brazil with headache, otalgia, runny nose, dysphonia, hyposmia, postural vertigo, weight loss of 5 kg and without visual changes. Rhinoscopy procedure was performed after clinical examination, which detected a “vegetating” lesion in the nasopharynx. A biopsy of the rhino pharynx tumor was then requested, with the removal of numerous irregular fragments of tissues with a light-brown surface, finely granular and fibroelastic, sent for anatomopathological analysis and initial imaging investigation with multislice computed tomography of the skull and Magnetic Resonance (MR) of the brain. The anatomopathological result suggested cartilaginous neoplasia with hypercellularity and nuclear hyperchromasia, concluding a histological aspect that could correspond to grade 1-2 chondrosarcoma depending on the correlation with clinical and radiological data. The multislice computed tomography of the skull (Figures 1 and 2) showed an expansive, heterogeneous and predominantly hypodense lytic lesion, with foci of calcification in the aspect of “popcorn” in-between, with irregular contours and defined limits, which presents a slight enhancement and predominantly peripheral after the infusion of the contrast medium, with an epicenter in the right petroclival synchondrosis. This formation invades the petrous region of the temporal bone and part of the sphenoid wing anterolaterally, without invading the orbital accumulation; extends medially and inferiorly through the jugular foramen, widening it, to the cerebellar point angle, right lateral wall of the cavum,

part of the clivus and without cleavage planes with the internal carotid artery, sphenoid sinus, and lateral retropharyngeal space, with consequent obliteration of the corresponding parapharyngeal space; it later compromises part of the cerebellum and the occipital condyle; finally, it affects the locoregional temporal lobe superiorly. The result of Brain Nuclear Magnetic Resonance Figure (3 and 4) suggested an expansive lesion whose epicenter is in the right petroclival synchondrosis, with erosion and bone destruction, characterized by an intermediate signal in T1, marked hypersignal in T2-weighted sequences, with some foci hypointense inside (calcification) and intense impregnation utilizing paramagnetic contrast. This lesion exhibits a component extending anteriorly and inferiorly to the retropharyngeal region and carotid space, maintaining close contact with the internal carotid artery and with the internal jugular vein, displacing the posterior wall of the rhino pharynx. Subsequently, the lesion occupies the cistern of the cerebellar-cerebellar and cerebellar-bulbar angles, causing an impression on the right anterolateral face of the bridge and bulb and the anterior face of the cerebellar hemisphere, extending further to the jugular foramen, petrous apex and part of the condyle occipital rights. Superiorly it extends to the parasellar region where it probably involves the cavernous sinus and circumferentially the internal carotid artery, also determining the impression on the medial aspect of the corresponding temporal lobe.

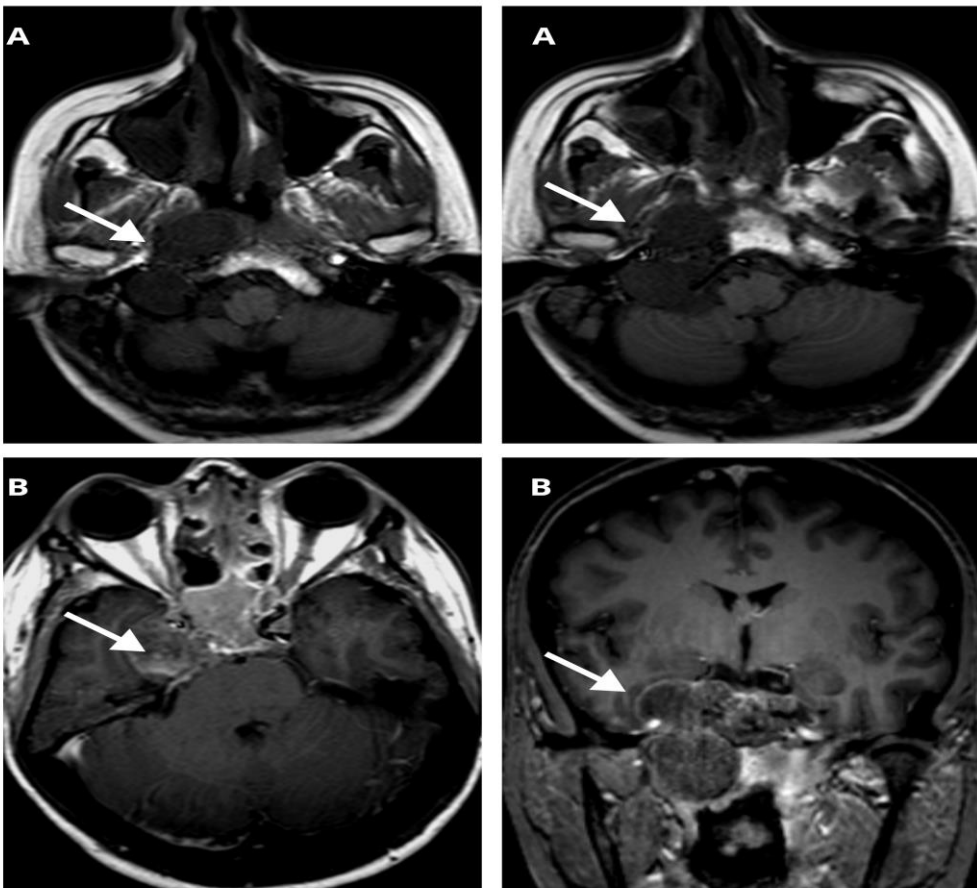


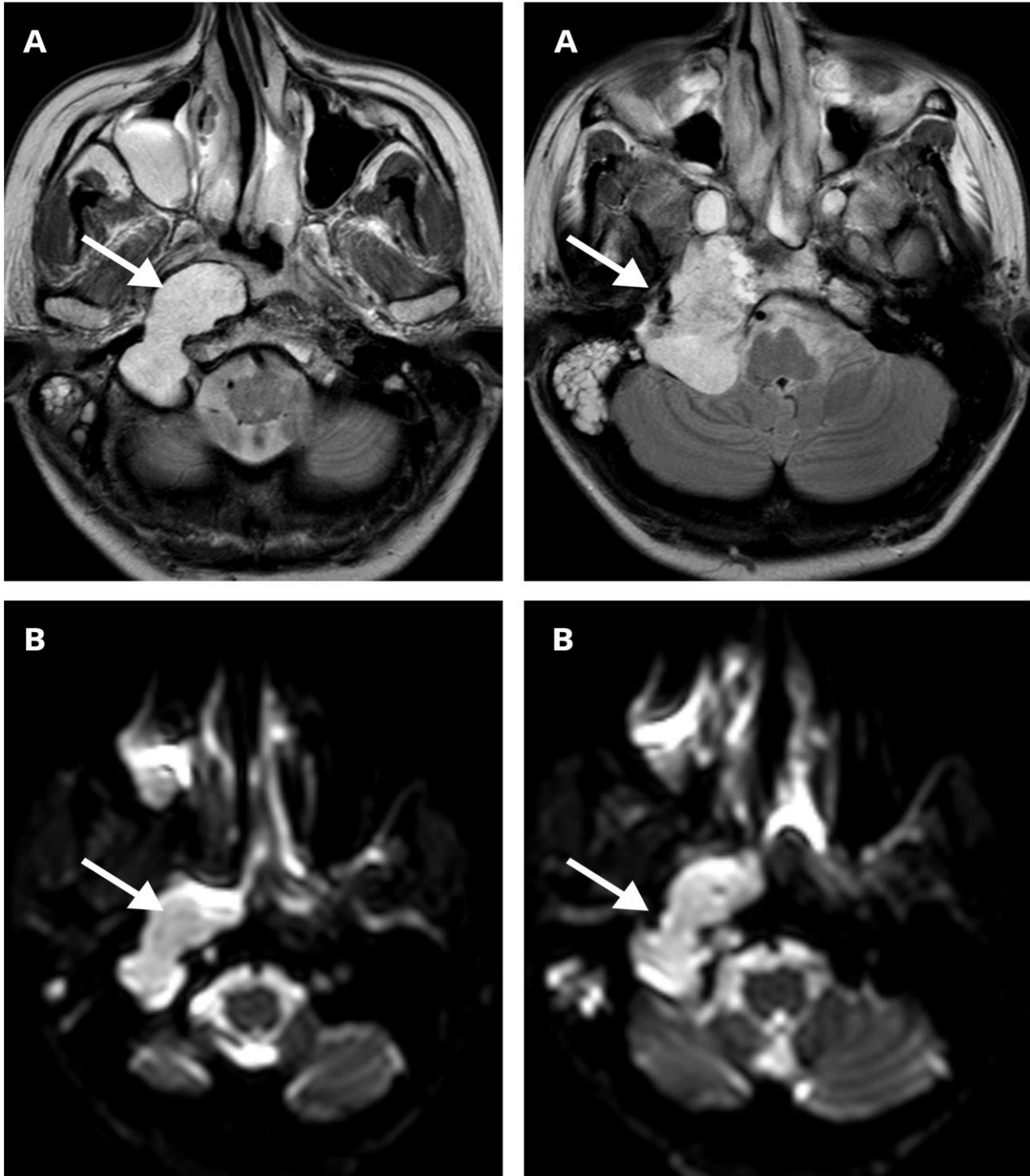
**Figure 1.** A. Axial CT scan using computed tomography, showing an expansive lytic lesion predominantly hypodense, with an epicenter in the right petroclival synchondrosis. B. Post-contrast axial and coronal section CT, showing slight and heterogeneous contrast to the contrast medium.



**Figure 2.** A, B- CT axial axial section in bone window, showing the lytic aspect of the lesion, and calcifications in “popcorn”.

**Figure 3.** A-axial T1-weighted MRI showing an intermediate sign of the expansive lesion in petroclival syndrosis. B- Axial and coronal MRI, weighted in T1 after contrast, showing enhancement by gadolinium.





**Figure 4.** A-axial T2-weighted MRI, showing a high sign of the referred lesion. B- Diffusion-weighted axial MRI showing a high signal in the ADC values.

#### DISCUSSION

Chondrosarcoma is a cancer of chondral origin, rare, slow-growing, locally advanced and with aggressive behavior, constituting 0.15% of all intracranial neoplasms, making up the third most common

cause of primary bone malignancy, after multiple myeloma and the osteosarcoma. Approximately 25% of all cranial chondrosarcomas occur at the base, representing 6% of all neoplasms at this site (1). This type of tumor has a slow growth, constituting locally



aggressive neoplasms that makeup 0.15% of all intracranial tumors, being capable of generating bone, cartilage or even tissues without cartilage constituents (1,2).

Embryology findings suggest the hypothesis that cranial-based chondrosarcomas may originate from multipotential mesenchymal cells or remnants of embryonic cartilage from cranial synchondrosis. The various plaques that remain after ossification of the chondroid fissures remain as growth sites for these tumors, and the most common tumor origin sites described were petroclival, petro-occipital, sphenoccephital, and sphenopetrous synchondroses, with a propensity for the first, that is, constituting parasagittal tumors in the majority. Other locations of expected impairment are the midline, more specifically in the basisphenoid/basioccipital, which are structures related to sphenoccipital cleft, and less commonly the junction between the nasal septum and the sphenoid face (1,3).

Histologically, chondrosarcomas are divided into conventional subtypes (myxoid and hyaline type), differentiated, clear cells and mesenchymal (1). The conventional form is the most common type at the base of the skull, being further subdivided into three classes: well-differentiated (grade I), moderately differentiated (grade II) and poorly differentiated (grade III) (1). The age range affected is variable; however, they often occur in middle-aged adults. The presentation depends on the location and the local extension; headache or paralysis of cranial nerves, particularly of the VI (abducent) nerve is a frequent sign. However, the patient, in this case, showed no signs and symptoms of visual impairment despite being diagnosed with petroclival chondrosarcoma (5).

As the petrous apex cannot be viewed directly, the radiological image plays a crucial role in the evaluation of injuries (4). Computed tomography (CT) presents a varied pattern, depending on the amount of the chondroid matrix. Generally, there are components of soft parts with a dense appearance in the non-contrast phase and enhanced by the

iodinated agent. Calcifications are characteristic, but not always present. A magnetic resonance imaging (MRI), these tumors usually present an intermediate signal at T1, a high signal at T2, with heterogeneous impregnation after the gadolinium injection (5,6). The main differential diagnoses include cholesteatoma of the petrous apex, calcified meningioma, chondromyxoid fibroma, chordoma, plasmacytoma, nasopharynx carcinoma, and metastases.

## CONCLUSION

Imaging examinations such as computed tomography and magnetic resonance become an important means of evaluation in this type of injury due to its difficult anatomical presentation. The indicated treatment combines surgery with several types of radiation; eventually, it consists exclusively of radiotherapy. Complete surgical excision is usually not feasible due to its location and proximity to neurovascular structures. However, as the growth pattern is slow, the prognosis is good (99% in 10 years). Although the VI cranial pair is affected in this type of injury, no characteristic visual symptoms were observed in the studied patient, highlighting the importance of the case report presented.

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