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Case report: Rare convexity meningeal chondroma mimicking a meningioma

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Case Report

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ABSTRACT

Background: Intracranial chondromas account for 0.2–0.3% of all intracranial neoplastic lesions and less than a quarter arise in the convexity or falx. Despite its benign nature, exceedingly rare malignant transformations exist. The misdiagnosis with meningiomas is frequent and may be related with chondromas' similar insidious clinical presentation and imaging features. Standalone surgery is advised and complete resection provides the definitive treatment.

Case Description: A 44-year-old female presents with insidious headache, visual disturbances, and papilledema. The imaging studies were compatible with frontal parasagittal meningioma. Surgery revealed a meningeal based mass, mostly avascular and with a well-demarked surgical plane from the brain parenchyma. Complete resection with meningeal margins was achieved and the histopathologic examination revealed a chondroma. The patient symptoms subsided and no surgical complications existed.

Conclusion: Intracranial convexity chondromas constitute a rare differential diagnosis for meningiomas. The present case reinforces the current scarce data and serves as reminder for clinicians diagnosing and treating intracranial tumors.

Keywords: Case report, Meningioma, Chondroma, Chondrosarcoma

INTRODUCTION

Intracranial chondromas (ICCs) account for 0.2–0.3% of all intracranial neoplastic lesions.^[9] These are usually located in the skull base and, in a recent review, only 6.7% of ICC arouse in the convexity.^[12] The etiopathology of ICC is not fully understood but speculations point to metaplastic fibroblasts or embryonal cartilaginous cells differentiation and proliferation resulting in a chondromyxoid matrix with mature chondrocytes.^[6,7]

ICCs have similar gender distribution and most are diagnosed between the second and fifth decades of age.^[4] Patients usually complain of headaches and diplopia.^[12] Seizures are not uncommon.^[12] Computerized tomography (CT) reveals well-demarked masses with variable density and slight to moderate contrast enhancement.^[3,13] Magnetic resonance imaging (MRI)

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hypointense T1 and hyperintense T2 sequences account for more than half of patients but diverse combinations exist. Most have calcifications, hyperostosis is frequent, and hemorrhage is rare.^[3,12] When occurring over the convexity, these clinical and imaging features raise an high suspicion for meningioma. Attempts to distinguish both pathologies may include an angiographic study where the almost avascular nature of chondromas is a distinctive clue.^[5]

Considered benign tumors, malignant transformation of ICC may occur in 2.3% of the cases. Most of the cases described were associated with Maffucci syndrome.^[12]

Surgical total resection of ICC is advisable. Incomplete resections frequently lead to reoccurrence and chemotherapy or radiation is not valid alternatives. Reoccurrence rarely follows complete resections.^[5,6,12]

CASE DESCRIPTION

A 44-year-old female with unremarkable medical history presents at our outpatients consultation with headache and visual disturbances described as "flashlights." Apart from an incipient papilledema, no other findings existed.

The CT scan revealed a left frontal parasagittal mass of 4 cm \times 4 cm \times 2 cm molding the surrounding brain parenchyma, mostly isodense, with a large central calcification and

hyperostosis [Figure 1]. The MRI reinforced the extraparenchymal nature of the lesion, revealing well-delimited margins and predominant hypointensity in T1-weighted images and mixed intensities in T2-weighted sequences. Furthermore, the mass effect seemed to compress and reduce the lumen of the superior sagittal sinus [Figure 1]. These findings enforce a diagnosis of meningioma. The lack of perilesional edema and the extra-axial noninfiltrative nature of the lesion made unlikely the diagnosis of a malignant tumor such as chondrosarcoma.

Surgical treatment was planned. Intraoperatively, a parasagittal mass pediculated from the convexity dura with lobular edges and a rubbery hard consistency was found. Its' well-demarked margins and defined dissecting plan from the parenchyma allowed for a complete *en bloc* removal with dural margins. No postoperative complications or sequelae existed and the previous symptoms subsided. The histopathologic analysis revealed a mass of cartilaginous tissue without cytologic atypia, confirming the diagnosis of dural chondroma [Figure 2].

DISCUSSION

The misdiagnosis of meningiomas in cases of chondromas is well described.^[11] Our patient symptoms were unspecific and probably related with the mass effect. The presenting



Figure 1: Convexity chondroma magnetic resonance imaging (MRI) and computerized tomography (CT) images. On the left and middle captions, T1- and T2-weighted MRI images reveal a dural attached convexity mass, well demarked from brain parenchyma and with no associated edema. No apparent invasion of calvarium diploe is noticed. On the right, a CT scan bone window highlights the calcifications inside the mass, responsible for the hypointense behavior in T1 and T2 MRI images.



Figure 2: Convexity chondroma on histology. Histopathological analysis reveals a monomorphous tumor of low cellularity. On the left and middle captions, we recognize chondrocytes without atypia embedded within a chondroid matrix. Immunohistochemically (on the right), neoplastic cells are S100 protein positive. In addition, they were vimentin positive and cytokeratin AE1/AE3, EMA, GFAP, and IDH1 negative (not shown). The Ki-67 labeling index was low (not shown).

symptoms, signs, and their evolution are similar in both pathologies.^[1,12] The onset in an younger patient raises the possibility of a chondroma and this fact is consistent with our case.^[12] Nonetheless, meningiomas can also occur in younger patients and their overall frequency is much higher.^[1,8]

When using CT studies, chondromas and most meningiomas appear as well-defined masses demarked from brain parenchyma. Furthermore, calcifications and hyperostosis are common in both cases.^[3,12,14] A gravitation toward chondromas may surge when the meningiomas' typical avid and homogenous contrast enhancement is absent^[3,11]. On MRI, chondromas may show several combinations of signal intensity and the signal intensity can be heterogeneous along the mass. All considered, no imaging finding is specific enough to differentiate chondromas from meningiomas^[12]. Our patient imaging features mimicked both chondroma and meningioma and the presumption of meningioma was due to its overall frequency. Features such as perilesional edema, diploe invasion, and less obvious hyperostosis were not present but these would raise the suspicion for a malignant lesion such as chondrosarcoma.^[10]

Almost all chondromas included in a recent systematic review followed surgical treatment.^[12] Furthermore, even when a diagnosis of meningioma is presumed, significant symptoms will prompt a surgical indication.^[1] Considering that chondromas may undergo malignant transformation and its reoccurrence is frequent in nontotal resections, efforts for a total resection should be pursued.^[2,12] Furthermore, no chemotherapy treatment exists for chondromas and radiation is both ineffective and may lead to malignant transformation.^[5,6] In our case, a total resection with margins respected all the above rationale.

In the histopathological analysis, the differential diagnosis of chondrosarcoma must be excluded. Increased cellularity, mitosis, atypia (nuclei are hyperchromatic and enlarged), and infiltrating borders are characteristic of chondrosarcomas and not compatible with chondromas.^[10,12] In our patient, we did not find these characteristics. The differentiation of chordomas from chondromas may also be important to take in mind, especially if you are facing a skull base lesion (which was not our case). The first has physaliphorous cells and shows pan-cytokeratin, EMA, S100 protein, and brachyury immunoreactivity.

CONCLUSION

Intracranial convexity chondromas are rare and can easily mimic meningiomas. The differential diagnosis is difficult or almost impossible before histopathologic confirmation. Our case underlines the need to keep this entity in mind since it will prone the decision to a surgical attitude due to its low but existent potential for malignancy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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