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Chondroma of the cerebral convexity: Case report

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Introduction: Intracranial chondromas comprise only about 0.2-0.5% of all intracranial tumors. It is considered slow-growing extra-axial benign tumors and may cause symptoms due to the mass effect on the cerebral parenchyma. The location of major incidence of these neoplasms is at the skull base, where it originates from residual embryonic chondrogenic cells. Very rarely, they can also originate from skull convexity, falx cerebri or cerebral parenchyma. We report a very rare case of an intracranial chondroma of the cerebral convexity.

Case report: 28 years-old female, with no previous comorbidities, sought medical assistance due to a progressive severe hemi-cranial headache. The neurological examination was unremarkable. Due to the progressive algic symptom, the patient underwent a magnetic resonance imaging (MRI) of the skull, which showed an extensive extra-axial lesion of 7.2x6.8x3.1cm in the left frontal lobe. The lesion showed heterogeneous signal intensity with flow voids within, but no significant contrast enhancement. The lesion determined a mass effect in the cerebral parenchyma, but there was no perilesional edema or abnormal signal. The hypothesis of intracranial chondroma was raised and neurosurgeons opted for surgical resection of the lesion due to the important mass effect and the risk of neurological deterioration.

During craniotomy, neurosurgeons noted an abnormal retraction of the dura mater in the corresponding area of the tumor. After dural opening, a hard whitish cartilaginous-like mass was identified. It was partially adhered to the dura and determined a compressive effect on the underlying brain. The tumor was slowly detached from the dura mater and removed en bloc. There were no signs of macroscopic invasion of brain tissue. Posterior histological analysis confirmed the diagnosis of intracranial chondroma.

The postoperative period was marked by a good recovery without the development of neurological deficits. The patient recovered well and remained stable clinically at last follows up.

Discussion: Intracranial chondromas of convexity are slow-growing tumors, large in presentation and may develop symptoms depending on the anatomical epicenter of the lesion, such as paresis, paresthesias, cranial nerves dysfunctions and seizures. The incidence of this disease has its peak in the third decade of life, usually between 20-60 years of age, with no gender predominance.

Chondromas arising from convexity or the falx is an extremely rare entity, comprising 20% of all intracranial chondromas. Accordingly with the most recent review of intracranial convexity chondromas, just 26 cases have been reported in the world, being our case the 27th patient with this condition. They can be found as solitary lesions or as part of Ollier's disease (multiple polysystemicenchondromatosis) or Maffuci's syndrome (multiple enchondromatosis associated with soft tissue angiomas).

Radiologically, intracranial chondromas are represented by a well-defined extra-axial lesion. In the computed tomography (CT), it may present as a hypo or hyperdense image, depending the degree of calcification. Also, it may present erosion/hyperostosis of the internal bone plate. In MRI, chondromas may present as a hypo or isointense on T1 and heterogeneous (hypo and hyperintense) on T2 with flow voids. In addition, cerebral edema is not observed and there is no contrast impregnation nor dural tail. Finally, in arteriography, chondromas are represented as an avascular area, which is a key point to the differential diagnosis with meningiomas (present tumor blush).

Several theories have been proposed to explain the origin of intracranial chondromas. Lesions located at the base of the skull are believed to originate from the residual embryonic chondrogenic cells along the basal synchondros is 6,7. However, for those who have their epicenter in cerebral convexity, falx cerebri or intraparequimatous, they may have their origin in the metaplasia of meningeal fibroblasts or perivascular mesenchymal tissue. In addition, some authors point to the heterotopic development of chondrocytes as a result of fibroblast activation by trauma or inflammation. Although all the discussion in the recent literature, the pathophysiology of this condition remains unclear.

These tumors do not respond to radiotherapy treatment, with a risk of inducing malignancy and are therefore not indicated even in the case of subtotal resection. Since intracranial chondromas are rare tumor entities, there are no grade 1 recommendations regarding their treatment. However, surgical treatment with complete removal of the lesion seems to be the best option especially because was no recurrence reported in all patients surgically treated with total resection.

Conclusion: Intracranial chondromas are extremely rare benign tumors that typically arise at the skull base and, in extremely rarer situations, may originate from the dura mater of the convexity. Most of these tumors have a good outcome when submitted to total resection of the lesion. Recurrence and malignancy rates are small and surgery is usually curative.