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Review of A Rare Malign Neoplasia: Clivus Chordoma

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Abstract

Background: Present a rare case of malignancy, correlate its different presentations and locations with its potential reserved outcome and review the possible imaging findings found in the assessment of brain structures. Summarize the main therapeutic possibilities.

Case report: We will report a case of 30 years-old patient, who discovery a clivus chordoma after entering the hospital with severe headache.

Conclusion: Imaging exams are decisive methods for diagnosing these tumors that grow slowly.

Keywords: Clivus Chordoma, Malign Neoplasia, Diagnostic Imaging, Radiology

Abbreviations: MRI: Magnetic resonance imaging

Introduction

Chordoma is a rare malignant neoplasm, originating from the embryonic remnants of the primitive notochord, with a prevalence of approximately 0.03 cases per 100,000 people in the United States [1], with variations in its location, but commonly found in the lumbosacral region and in cerebral clivus, presenting itself as a surgical challenge due to the proximity to vital neurovascular structures and the high potential for recurrence [2]. About 25% of chordomas originate in the clivus and constitute approximately 0.15% of all primary intracranial lesions [3], corresponding to 1 to 4% of all malignant bone tumors. Microscopically, chordoma is a moderately cellular neoplasm composed of vacuolated *physaliferous* cells arranged singly and in cords within a myxoid stroma [4].

A high percentage of incomplete resections is observed in the literature, due to the characteristics of high malignancy and tumor recurrence, as well as the involvement of irresistible

structures. It presents itself as a therapeutic possibility, radiotherapy and, in selected cases, endoscopic transsphenoidal endoscopic surgery, which allows the approach of a minimally invasive form of expansive lesions of the skull base [2]. The prognosis of the disease is reserved.

Case Report

A 30-year-old patient undergoes an elective consultation with the neurologist, reporting a progressive headache associated with dizziness and asthenia; before the reassessment with the specialist, the patient comes to the emergency department, with severe headache, and the MRI exam service was performed, observing a large expansive formation. The patient underwent surgical resection for biopsy, with no possibility of complete extraction due to intrinsic and irresistible characteristics of the tumor. Anatomopathological examination confirmed that it was a chordoma of the clivus.

Patient evolves with visual alteration and paralysis of the extraocular muscles. The patient is currently undergoing chemotherapy and radiation therapy.

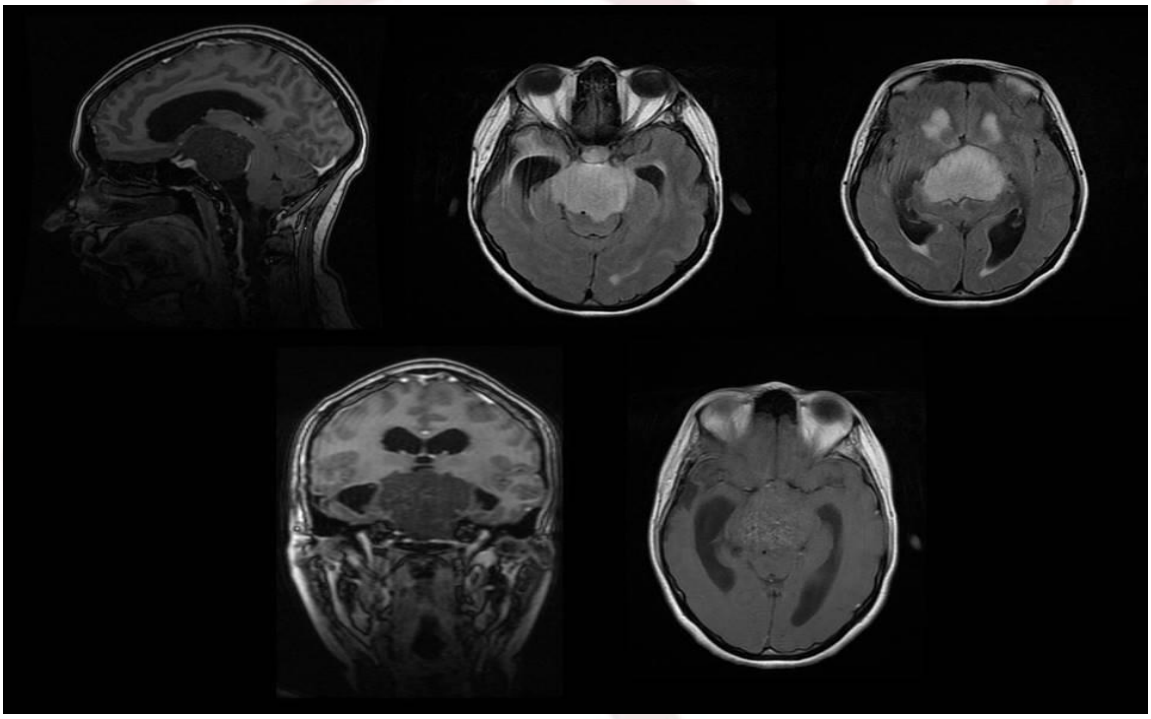

Fig 1: In T1 sagittal section, a massive, extra-axial, hypointense expansive formation with a mass effect is observed, located in the suprasellar region. In the FLAIR power, axial cuts, a lesion with a hypersignal is observed, which displaces and compresses the optic chiasm, hypothalamus and later the midbrain and bridge. In coronal section T1, tumoral lesion with hyposignal, of large dimensions, centered and suprasellar, which compresses the third ventricle, associated with hydrocephalus and diffuse deletion of the cortical grooves. Discrete and heterogeneous focal captures by means of contrast in the post-contrast image.

Discussion and Conclusion

Voluminous expansive formation of approximately 4.6 x 4.4 x 4.3 cm, centered in the extra-axial compartment of the suprasellar region, perimesencephalic, pre-pontine and hypothalamic, predominantly hypointense in T1, hyperintense in T2 and with foci of discrete and heterogeneous signal by means of contrast.

The tumor compressed and displaced the optic chiasm, the hypothalamus and the III ventricle superiorly, producing important hypertensive hydrocephalus as well as displacing and subsequently compressing the midbrain and bridge, suggesting neoplasia of the supraseal region, suggesting the possibility of craniopharyngoma initially.

Chordomas are rare and slow-growing tumors, with a strong tendency to local invasion and recurrence, however metastases are uncommon. Imaging methods play a decisive role in the evaluation, determination of the characteristics and extent of the tumor, being fundamental for diagnostic thinking and in the correlation with the differential diagnosis. Early diagnosis and treatment are essential measures to increase the disease's survival.

Authors' contributions

AJ designed the study, acquired and interpreted the data, and have to be personally accountable for the accuracy and integrity of the entire work. NA and PC provided clinical care to the patient, performed literature searches, interpreted the data, and drafted the manuscript. LG, JR and JG collected, analyzed, interpreted the data, study, conception and patient consent. All authors reviewed and revised the manuscript and approved the final manuscript.

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