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# Intradural chordoma presenting with intratumoral bleeding

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ABSTRACT

Intradural clival chordomas are very rare, and only 29 cases have been reported to our knowledge. They arise purely intradurally without bone or dural involvement and may differ from classic clival chordomas in physiopathology and management. We present a 28-year-old woman who presented with intradural clival chordoma and tumoral bleeding. After initial gross macroscopic surgical resection, she presented with tumor recurrence after 2 years, again with intratumoral bleeding. Although usually considered to have a more favorable prognosis in comparison to typical chordomas, intradural chordomas appear to behave as typical chordomas. Intratumoral bleeding may be a sign of an aggressive lesion and risk of recurrence. We highlight the differential diagnosis of intrinsic posterior fossa bleeding, especially in young patients. Intradural chordomas may be underdiagnosed and incorrectly treated as other types of parenchymal hemorrhage.



## 1. Introduction

Chordomas are rare intracranial neoplasms which originate from remnants of the primitive notochord, with origin of approximately 35% in the clivus and 50% in the sacrococcygeal region. Chordomas account for 1% of all primary brain tumors [1–4].

Intradural clival chordomas are rare, and only 29 cases have been reported to our knowledge [1]. They arise purely intradurally without bone or dural involvement and may differ from classic clival chordomas in physiopathology and management [5–8]. We present a patient with an intradural clival chordoma presenting with tumoral bleeding and review the pertinent literature.

## 2. Case report

A 28-year-old woman with a previously unremarkable medical history presented to the emergency department with sudden-onset headache and dizziness in November 2012. At admission, neurologic examination revealed a Glasgow Coma Scale score of 15, reactive pupils, and right-sided hemiparesis (grade III in right upper extremity, grade IV in right lower extremity). Additionally, she had left facial numbness and a left abducens nerve palsy. No other cranial nerves were involved.

CT scan of the head revealed extensive bleeding anterior to the brainstem, predominantly on the left side, extending from the mesencephalon to the medulla (Fig. 1). She was initially managed as a spontaneous intracranial hemorrhage in a young adult, with a thorough laboratory, endocrine, and rheumatologic work-up. Vascular, hematologic, and rheumatologic causes were excluded. She also underwent MR angiography, which confirmed the bleeding as well as gadolinium enhancement. Diffusion-weighted images revealed hyperintensity.

After 1 month of neurological observation and exclusion of other causes, she was discharged to outpatient follow-up with normal strength and complete resolution of the abducens nerve palsy. One month after discharge, however, the patient experienced another episode of headache with dizziness, and returned to the emergency department. On neurologic

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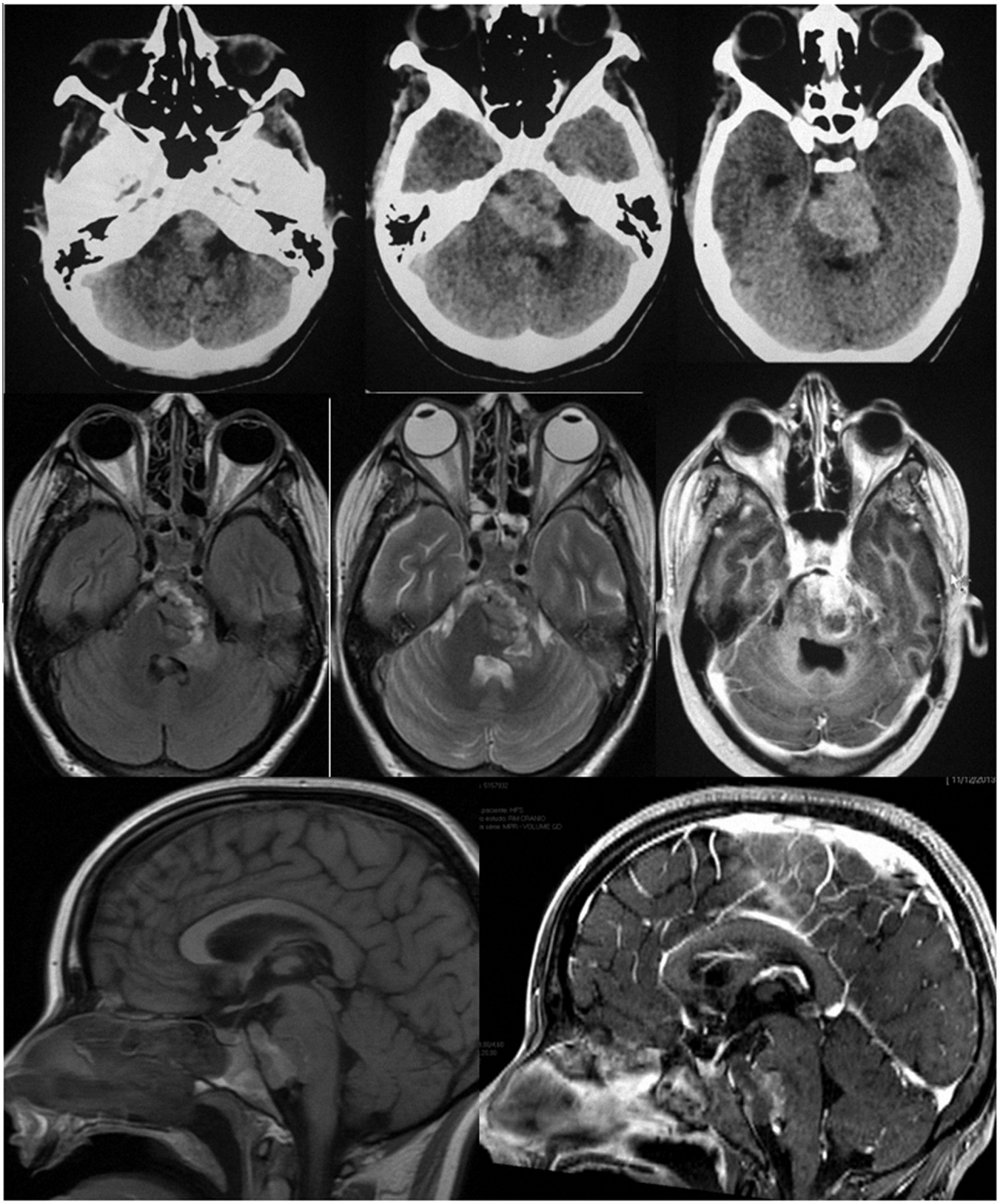

**Fig. 1.** (Top) Axial CT scans at presentation showing a posterior fossa hematoma. (Middle) Axial MRI with fluid attenuated inversion recovery (left), T2-weighted (middle) and T1-weighted with contrast sequences showing the brainstem intradural chordoma. (Bottom) Sagittal T1-weighted MRI with (right) and without (left) contrast showing the brainstem intradural chordoma, arising from the clival synchondrosis, which strongly suggests the diagnosis of chordoma.

examination, she was comatose, and a new hematoma was detected. Emergent surgical evacuation of the brainstem hematoma was performed via a retrosigmoid approach, followed by a presigmoid approach 1 week later, with final complete resection of the tumor. Pathologic examination confirmed an intradural chordoma (Fig. 2).