



Pediatric Clival Chordoma Presenting as Nasal Congestion and Sleep Disordered Breathing

Cedars
Sinai

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Key Points

- Pediatric clival chordoma may present with atypical symptoms that mimic common ENT conditions
- Advanced imaging techniques, including CT and MRI, are crucial for diagnosing clival chordomas
- Surgical resection is the primary treatment modality with close surveillance postoperatively

Introduction

- Chordomas are rare malignant tumors derived from notochord remnants, developing at various points along the craniospinal axis^{1,2}
- The incidence is estimated at 8 per 10 million individuals, with most diagnoses occurring in the fourth decade of life¹
- In children, chordomas are exceedingly rare, constituting only 5% of cases
- They often extend intracranially, commonly affecting the clivus^{1,2}
- Chordomas are slow-growing yet invasive, frequently remaining undetected until they reach advanced stages
- Common symptoms include headaches, diplopia, and cranial nerve deficits^{2,3}
- While there are few documented cases of pediatric clival chordomas, extrasosseous tumor causing nasal obstruction as the primary symptom is even more rare
- We report a patient who presented with nasal congestion and sleep-disordered breathing, initially believed to have a Thornwaldt cyst, but determined to be a clival chordoma upon further workup

Case Presentation

- 12-year-old male was referred for six months of persistent nasal obstruction and loud snoring
- Presented with chronic nasal congestion and mouth breathing, along with daytime sleep disturbance symptoms
- Treated previously with oral antihistamines and intranasal steroids
- Physical exam revealed tonsillar and turbinate hypertrophy with notably smooth adenoids
- Mild improvement after two months of daily nasal steroid and antihistamine therapy
- Due to the unusual adenoid appearance, CT scan was obtained demonstrating sinus disease and a cystic nasopharyngeal mass extending from the clivus
- MRI confirmed no intracranial involvement (Figure 1).
- Differential diagnosis included a Thornwaldt cyst and nasopharyngeal chordoma, both originating from notochordal remnants
- Nasal endoscopy with biopsy was performed (Figure 2) revealing chordoma, and subsequent imaging showed attachment to the ventral clivus
- Tumor board consensus recommended surgical resection as the primary treatment
- Intraoperatively, fibrotic changes along the dura were identified and managed with bipolar cautery, and large skull base defect was repaired with a nasoseptal flap
- No immediate postoperative complications and patient was discharged POD #1
- Surveillance MRI POM #8 indicated residual versus persistent tumor along the dorsum sellae leading to a second resection.
- Adjuvant proton therapy decided against after considering the risk of pituitary gland radiation in our young patient

Figures

Pre-Operative MRI

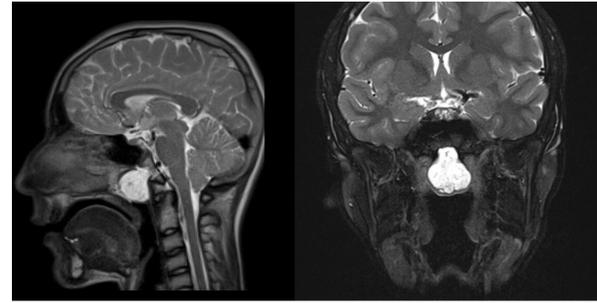


Figure 1. T2-weighted MRI sinus with contrast (sagittal - left; coronal - right)

Nasal Endoscopy



Figure 2. Initial nasal endoscopy and biopsy

Histopathology

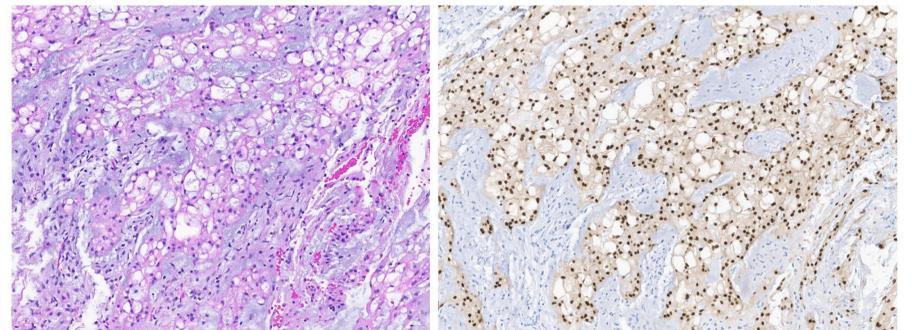


Figure 3. Frozen section histology (Hematoxylin and eosin stain - left; Brachyury immunohistochemistry - right)

Post-Operative MRI

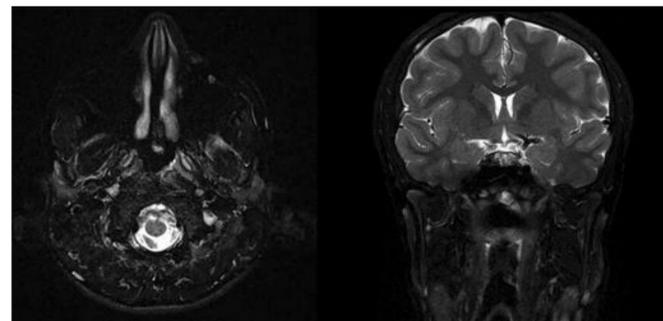


Figure 4. T2-weighted fat saturated MRI sinus (axial - left; coronal - right)

Conclusions

- Pediatric clival chordomas, though rare, pose significant clinical challenges due to subtle presentation, similarity to benign pathology, and lack of consensus on management
- Case illustrates the importance of considering chordoma in children with atypical symptoms such as nasal congestion and sleep disturbances
- Biopsy should be considered in any suspicious nasopharyngeal mass, especially with concomitant bony erosion on imaging
- Case aids in refining diagnostic and therapeutic approaches and contributes to the growing body of literature on pediatric clival chordomas

Contact

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