Minimally Invasive Resection of Pediatric Clival Chordoma Using Ultrasound Guidance

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Introduction

Clival chordomas are rare, slow-growing malignant tumors that arise from notochordal remnants at the skull base. They represent some of the most challenging skull base tumors to treat because of their **locally invasive nature**, **high rates of recurrence, and associated mortality.**[1] Patients may present with diplopia, blurred vision, facial numbness, or headaches. Although skull base chordomas most commonly affect older adults, they can rarely occur in **pediatric patients**, in whom the disease may exhibit **a more aggressive clinical course.**[1]

Surgical management of clival chordomas is particularly complex due to their **deep-seated location and proximity to critical neurovascular structures**. The clivus is bounded superiorly by the dorsum sellae, laterally by the cavernous and paraclival segments of the internal carotid arteries, inferiorly by the foramen magnum, and posteriorly by the brainstem. These anatomic constraints often limit the ability to achieve complete resection, as **tumors frequently abut or encase the carotid arteries and lower cranial nerves**. While gross total resection is essential for durable disease control, it carries substantial risk given the close relationship to these vital structures. In this context, **intraoperative ultrasound has emerged as a valuable adjunct, providing real-time visualization of vascular anatomy**—particularly the carotid arteries—during endoscopic skull base surgery.

Case Presentation

An **18-year-old male** presented to the otolaryngology clinic with **recurrent epistaxis.** Nasal endoscopy was performed and demonstrated a vascular mass filling the right nasal cavity. An MRI was obtained to evaluate the mass and incidentally demonstrated an enhancing lesion involving the inferior third of the clivus. This second mass extended anteriorly to the cranio-cervical junction and occupied the space between the clivus and the odontoid process. The lesion was closely associated with the paraclival carotid arteries and abutted the brainstem.

Procedures

The patient initially underwent resection of the vascular nasal tumor emanating from the right middle turbinate. Subsequently, a **minimally invasive endoscopic endonasal approach** was used to address the clival lesion. Intraoperative biopsy confirmed the diagnosis of chordoma.

Intraoperative ultrasound was employed to identify the **bilateral carotid arteries**, allowing for precise dissection and reducing the risk of vascular injury. Notably, the **resection was accomplished without division of the palate or posterior septectomy**, preserving normal anatomy and reducing morbidity.

Gross total resection of the chordoma was achieved, as confirmed by postoperative MRI. The patient received adjuvant radiation therapy and **remains disease free after 18 months.**

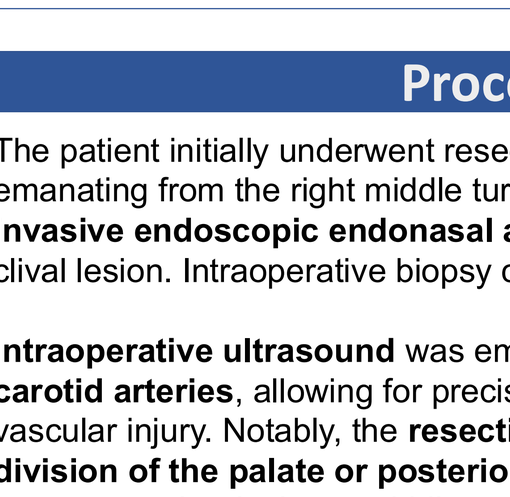

Figure 1. Pre-operative MRI scan of clival lesion that was incidentally found.

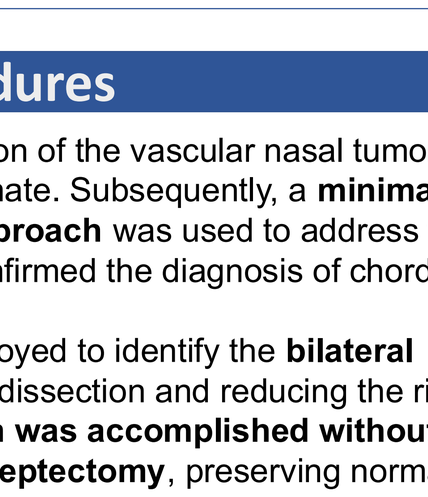

Figure 2. Post-operative MRI scan showing the gross total resection of the clival lesion.

Discussion

This pediatric case offers a unique perspective given its unusual clinical presentation, abnormal radiological features, and unique minimally invasive approach aided by novel ultrasound technology during resection.

Unusual clinical presentation

The presentation of this case was **uncommon** given the **clival lesion was incidentally revealed during an MRI** of a rare vascular mass of the right nasal cavity for a primary complaint of epistaxis. There is **no published case report with a similar presentation** of a chordoma concurrently with a vascular nasal tumor.

Abnormal radiological features

This case illustrates **the diagnostic complexity of pediatric clival chordomas.** Even though the chordoma was T1 isointense and T2 hyperintense, the **lesion did not have other typical features of chordomas**, namely a lobulated appearance and locally destructive features.

Unique minimally invasive approach

In recent years, the use of endoscopic endonasal approaches in pediatric skull base cases has increased significantly due to **its higher resection rate and lower surgical complications.**[1,2,3] Our approach was unique as it was completed **without the need for posterior septectomy, sphenoidotomy, posterior ethmoidectomy, or palate division.**

Novel use of ultrasound technology

This case involved the novel use of intraoperative ultrasound to **enhance the safety of the minimally invasive endoscopic approach** through dynamic, real-time identification of the skull base vasculature, including the carotid arteries. Better visualization between the tumor and vascular structures **aided a gross total resection of the mass**. It also improved the surgeons' ability to assess the extent of the tumor during resection.

Limitations of ultrasound technology is the **absence of a three-dimensional view** to help surgeons identify and visualize structures.[3] As this technology grows in prevalence, it will be important to address these issues.

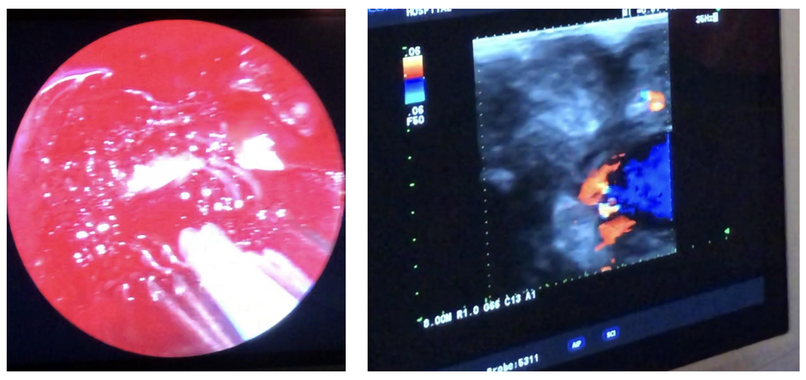

Figure 3. Use of intraoperative ultrasound during endoscopic skull base surgery.

Conclusion

This pediatric case not only **illustrates the complexity of pediatric clival chordoma resection, but also offers a unique perspective** given its unusual clinical presentation, abnormal radiological features, and unique minimally invasive approach.

The use of **intraoperative ultrasound** enabled dynamic, real-time identification of the carotid arteries, enhancing the safety of the minimally invasive endoscopic approach. Importantly, the procedure was completed without the need for division of the palate or a posterior septectomy.

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References

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