

Introduction

- Clival chordomas are rare, locally aggressive tumors arising in the clivus from notochord remnants.
- Patients typically present with cranial nerve palsies including diplopia, headaches, and dysphagia.
- These lesions typically present in individuals age 40-60 years of age with a male predominance.
- Treatment includes surgical resection followed by adjuvant radiation.

Case Report

- A 37-year-old male presented to the ED with chief complaint of neck pain, hoarseness and dysphagia for the past two days after playing volleyball.
- Flexible laryngoscopy was performed demonstrating complete right vocal cord paralysis.
- CT was obtained which demonstrated a destructive mass centered right of midline in the posterior skull base and upper cervical spine with deformity of the lower brainstem and upper cervical spinal cord. Chronic denervation of cranial nerves IX through XII on the right.
- MRI demonstrated large lytic skull base lesion involving clivus and petrous apex with extension into the right hypoglossal canal and jugular foramen. Differential included chondrosarcoma versus chordoma.
- Prior to resection, he underwent biopsy of the lesion, tracheostomy and G-tube placement. Biopsy results demonstrated chordoma.
- Prior to tumor removal, the patient underwent a posterior cranial cervical fusion (occiput to C2).
- Tumor was removed with collaboration between neurosurgery and ENT in both an endoscopic transnasal and transoral fashion.
- The inferior aspect of the tumor was exposed by dividing the soft palate. This allowed for visualization of pre-vertebral musculature which was divided and reflected laterally (Figure 3).
- A gross total resection was achieved, and reconstruction was performed by our head and neck colleagues.
- Patient also underwent right injection medialization laryngoplasty due to paresis of right vocal cord and subsequent aspiration.
- Reconstruction was performed with an anterolateral thigh free flap due to the need for adjuvant radiation and high risk of CSF leak given the location of the tumor and the difficulty in securing grafts in the posterior fossa.
- Free tissue transfer is also advantageous in preventing osteomyelitis following radiation.

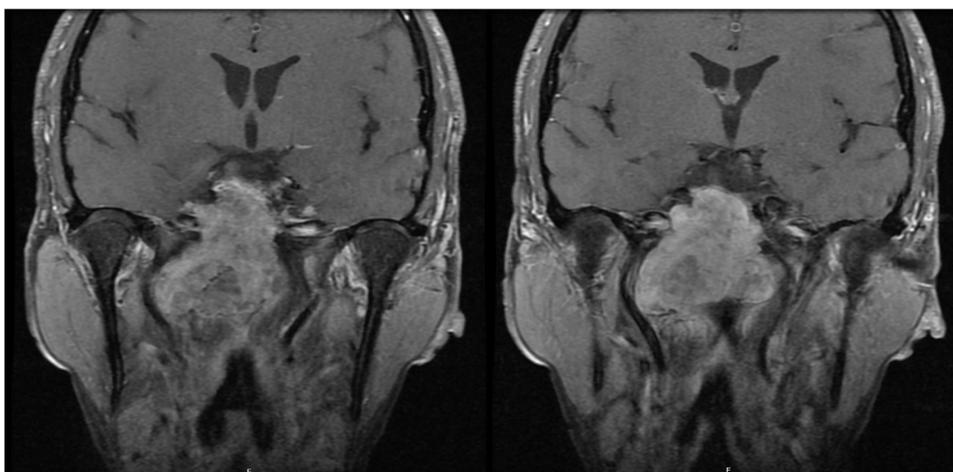


Figure 1: MRI demonstrating large lytic bone lesion involving clivus and petrous apex with extension into hypoglossal canal and jugular foramen.

- The location of the chordoma and clival invasion required dissection inferiorly to the nasopharynx and pharynx which significantly affected his swallowing function. Moreover, some of the patient's swallowing function was diminished from the tumor involving lower cranial nerves.
- For this reason, the patient also underwent pharyngoplasty to improve his swallowing function post-operatively.

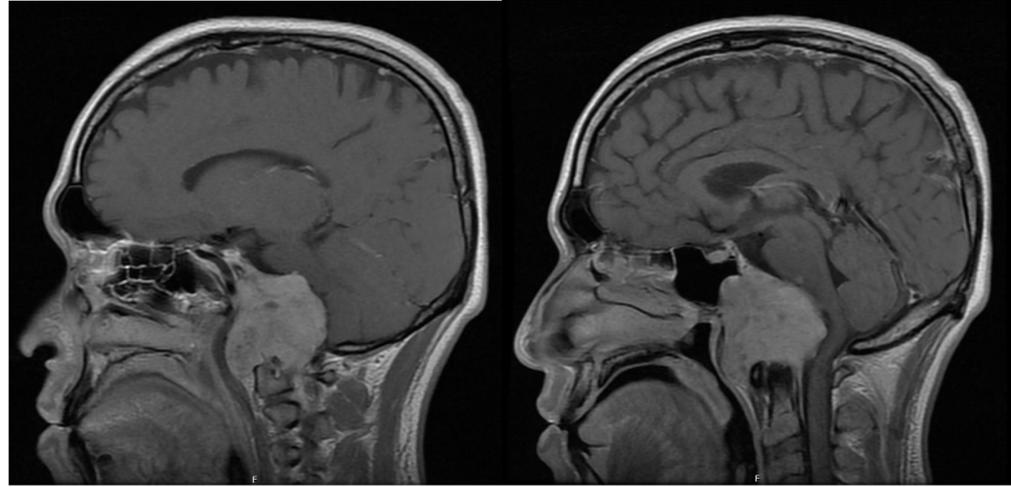


Figure 2: MRI demonstrating large clival chordoma with compression of lower brain stem and upper cervical spinal cord.

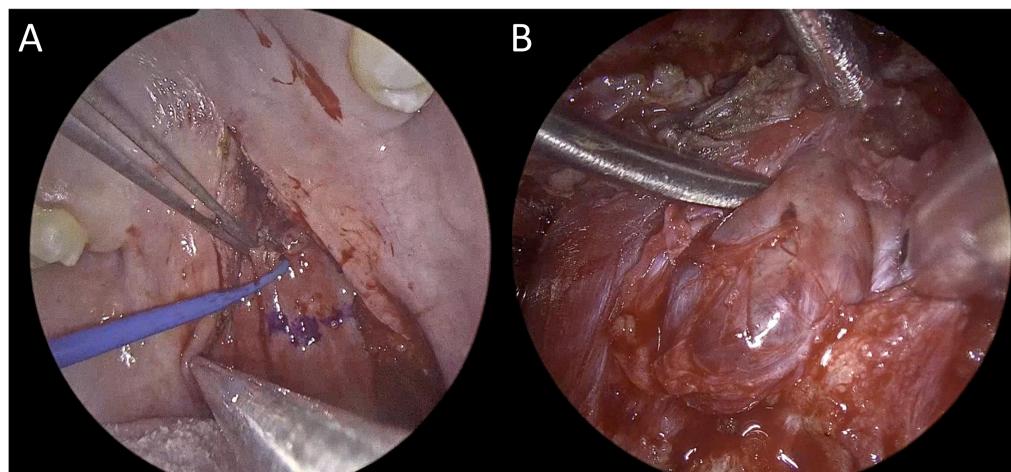


Figure 3A & 3B: A: Intraoperative photo depicting division of soft palate for inferior exposure. B: Photo of tumor with endoscopic dissection being performed.

Conclusions

- Clival chordomas are rare, slow growing tumors with a male predominance.
- Presentation of these tumors usually includes cranial nerve deficits commonly involving the lower cranial nerves due to their proximity to the clivus.
- Pre-operative work up with CT and MRI are essential for surgical planning.
- Removal and management of these tumors and the sequelae associated with these tumors is best accomplished with a multi-disciplinary team including neurosurgery, rhinology, and head and neck oncology.
- Vascularized flaps are an excellent option for reconstruction as most clival chordomas require post operative radiation due to high recurrence rates.

References

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