

Headmirror's ENT in a Nutshell

Clival Chordoma

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Presentation (0:38)

- Symptomatology
 - o Diplopia (injury or stretching of abducens nerve as it passes through Dorello's canal)
 - o Headache
 - o Facial numbness from trigeminal numbness
 - o Vague symptoms
- Epidemiology
 - o Rare tumor (1:1,000,000)
 - o Late to middle age
 - o No risk factors identified
- Differential diagnosis
 - o Chondrosarcoma
 - o Lymphomas
 - o Adenocarcinomas
 - o Bony dysplasia
 - o Nasopharyngeal mucosal tumor with extension into the clivus

Relevant Anatomy (1:56)

- Fusion bone at base of skull (part of occiput and part of sphenoid)
 - o Back of oropharynx and nasopharynx
- Upper 1/3: Clinoid to floor of the sella
- Middle 1/3: Floor of the sella to floor of sphenoid sinus
- Lower 1/3: Floor of the sphenoid down behind the nasopharynx

Pathophysiology (3:58)

- Chordoma is derived from notochord remnant (spans from sacrum to clivus)
- Physaliferous cells (soap bubble appearance on histopathology)
- Brachyurea positive, distinguishes from other tumor
- 1/3 of all chordomas are in the clivus
- *Is this a malignancy:* yes, locally progressive, can metastasize and cause mortality
- Subtypes
 - o Classic (conventional chordoma)
 - o Chondroid chordoma
 - o De-differentiated (atypical chordoma): higher mitotic count, more aggressive tumors

Workup (7:16)

- Imaging
 - o CT: locally destructive lesion in the clivus, typically in the midline

- Chondrosarcoma can have deposition of calcium in them
 - MRI: hyperintensity on T2 signal (chondrosarcoma enhance on T2 as well) and contrast enhance
- Biopsy: do not biopsy in clinic because it can seed
- MR negative resection
 - Negative MRI prior to proton therapy is an indicator of survival

Treatment (10:35)

- Surgical Approach
 - Endoscopic endonasal expanded approach
 - Raise unilateral large extended nasoseptal flap preserving it in the maxillary sinus during resection
 - Contralateral nasoseptal flap will be raised as well
 - Large sphenoidotomy, open up to the planum, and posterior septectomy
 - Drill down floor of sphenoid sinus to get flushed to the clivus
 - Vidian nerve traced back to the carotid artery
 - Exposure from sella to the lower 1/3 of the clivus
- Radiation Therapy
 - Proton beam (some centers still use IMRT and gamma-knife)
 - Can achieve high dose adjacent to the brain stem
- Brachyurea
 - Increases sensitivity to radiation
 - Targeted therapy (future directions)
- Follow up
 - Post operative debridements initially
 - MRI 3 month after followed by every 6 months for the first few years
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