Headmirror's ENT in a Nutshell
Esthesioneuroblastoma
Experts: Garret Choby, M.D. and Jamie Van Gompel, M.D.

Presentation (0:28)

- Symptomatology
  - Nasal obstruction
  - Hyposmia or anosmia
  - Epistaxis
  - Headache, vision changes
  - Rare presentation cervical metastasis (5% patients)
- Physical Examination
  - High nasal vault tumor (olfactory cleft)
  - Fleshy mass, moderately vascular
  - Thorough cranial nerve and neck examination
- Epidemiology
  - Bimodal age distribution
  - SEER database → middle to late age (40-60s)
- Differential Diagnosis
  - Squamous cell carcinoma
  - Sinonasal undifferentiated carcinoma
  - Sinonasal neuroendocrine carcinoma
  - Lymphoma (imaging can look very similar)
  - Rhabdomyosarcoma
  - Ewing Sarcoma
  - Sinonasal mucosal melanoma

Pathophysiology (2:20)

- Theorized to arise from bipolar cell from nasal epithelium (upper 1.5 cm of the nasal vault)
- Small blue round cell tumor or neuroendocrine tumor
- Homer-Wright rosettes (1/3 of patients)
- Flexner wintersteiner rosettes

Workup (6:00)

- Imaging
  - CT scan
    - Boney erosion at the skull base
  - MRI
    - Uniformly enhancing mass in the upper nasal vault with possible extension into the paranasal sinuses and intracranially
    - Dumbbell appearance is not reliable sign clincally
  - PET-CT scan
    - Especially for Kadish C-D lesions

        - Metastatic work up usually not performed for small lesions or Hyams grade 1-2
- <u>Biopsy</u>
    - Most important next step after imaging, especially in clinic

- <u>Grading System</u>
    - **Kadish Staging System**
        - A: confined into the nasal cavity
        - B: extension into the paranasal sinuses
        - C: tumor beyond nasal cavity and paranasal sinus which can include cribiform plate, erosion of skull base, intracranial cavity or orbital invasion
        - D: cervical nodes or distant metastasis
    - **Hyams Grading System**
        - Can be applied to many adenocarcinomas
        - Hyams grading is an independent risk factor for these tumors being aggressive
        - Hyams grade 1-2 (low grade) lesion compared to 3-4 (high grade)

Treatment (12:45)
- Treatment modalities are controversial
    - Upfront surgical resection if margin negative resection can be achieved at our institution
    - Other institutions favor upfront neoadjuvant therapy
- Localized tumors
    - Endoscopic endonasal resection
        - Exposure will be orbit to orbit with Draf III frontal sinusotomy and open up both sphenoids widely
        - Bony osteomies with a drill
    - Majority of cases can be treated with these
    - If negative margins cannot be achieved, convert to craniofacial resection
    - Multi-layered reconstruction with fascia lata graft as in-lay(possible Duragen above that) and extended nasoseptal flap on-lay
        - If NSF not available, dual layer fascia lata graft in-lay and on-lay is used
- Post-operative radiation
    - Most radiation literature support adjuvant radiation
    - At our institution, with low Hyams grade and margin negative resection, we will give the patient option for no radiation
        - Those patients without radiation have higher incidence of recurrence, but surgical salvage is an option
- Chemotherapy
    - Reserved for metastatic disease
    - No adjuvant therapy for postoperative radiation (only neoadjuvant for pre-op)
    - No data that any agents radiosensitizes these tumors

    - Neo-adjuvant may be used up front to shrink tumor to achieve margin negative resection
- Neck Management
    - Elective neck dissection not recommended
    - High-rate of delayed neck metastasis (15%)
        - Salvage neck dissection vs radiation therapy
    - Some centers may perform elective neck radiation (controversial)
    - Having a single neck node is #1 predictor of mortality

Prognosis (22:55)

- Median survival 8.5 years
    - Hyams grade, Kadish stage, margin negative resection important factors
- SEER database
    - Hyams grade 1-2: 66% 10 year survival
    - Hyams grade 3-4: 30-35% 10 year survival

Follow Up (24:14)

- Scan at 3 months postoperatively followed by every 6 months for the first few years
- Nasal endoscopy important for surveillance as well as clinical neck examination
- Do not typically follow them with PET-CT