

Esthesioneuroblastoma (olfactory neuroblastoma): Read the summary in the brown book as well

- Uncommon tumour of the upper nasal cavity, probably of neural crest origin with neuronal and epithelial differentiation 1-5% of malignant nasal tumours.
- Histologically composed of small round or oval cells arranged in lobules among neurofibrillary background. Rosettes and Pseudorosettes are occasionally found. Cells stain +ve for Synaptophysin, chromogranin A, cytokeratin and EMA. Ki-67: 0-40%. Electron microscopy-secretory granules
- Presentation: epistaxis, nasal obstruction, anosmia, occasionally with increased ICP in case of large intradural lesion. Reported cases with SIADH and Cushing's syndrome. Metastases usually occur first to cervical lymph nodes in up to 48%. Distant metastases to bone, bone marrow, lung and skin and leptomeningeal metastases are associated with poor prognosis. Peak incidence 10-20 and 40-50 years.
- Differential diagnosis: olfactory groove meningioma, nasopharyngeal carcinoma, cystic adenoid carcinoma, rhabdomyosarcoma, juvenile angiofibroma, melanoma
- Investigations: CT scan axial and coronal with bone windows, MRI- Tumour with epicentre under the cribriform plate with homogenous enhancement, Nasal endoscopy with biopsy is diagnostic, isotope scan with radioactive octreotide can detect metastases (**most tumours express somatostatin receptors**)
- Grading :
 1. Clinical : kadish stage 1 (nasal), 2 (paranasal sinuses), 3(intracranial or orbital), 4 distant metastasis
 2. Pathological (Hyman's):4 grades depending on the presence of hypercellularity, pleomorphism, mitosis and necrosis
- Treatment:
 1. Craniofacial resection (lateral rhinotomy and frontal craniotomy) with attempted gross total resection+ radical neck dissection in the case of cervical Mets. If the tumour margin is free and the lesion is low grade follow up by MRI every year for 5 years and then every other year indefinitely
 2. Postoperative radiotherapy for residual and high grade tumours. Some centres give preoperative radiotherapy

3. Preoperative chemotherapy for large tumours using cisplatin and etoposide and for systemic metastases
 - Median 5 year survival is 70% and may be affected by the tumour's stage, grade, presence of metastases and the patients' age. Follow up should be life long because these tumours can reoccur many years after resection (40% 5 years or more).