

## A BRIEF REVIEW OF HEAD & NECK PATHOLOGY

Although there are many pathologic entities peculiar to the head and neck area, a variety of bone, soft tissue, and epithelial lesions which are not site specific are also seen there. The lesions discussed here include both common and uncommon entities and are presented for their differential diagnostic relevance or for their uniqueness in location or clinical presentation. Most of the discussion will revolve around clinical presentation.

### Two Life-Threatening Infections

Necrotizing External Otitis  
Rhinocerebral Phycomycosis

#### 1. Necrotizing External Otitis:

- Potentially lethal, rapidly progressive external otitis due to *Pseudomonas aeruginosa*.
- Seen in older patients with some immune compromise: diabetes, chronic debilitation, immunodeficiency disorders.
- Begins in external auditory canal with minor trauma, progresses with pain swelling, granulation tissue, purulent otorrhea.
- Advanced stage with osteomyelitis, chondritis, extension to skull base and middle ear space.
- Intracranial extension characterized by multiple cranial nerve palsies, meningitis, venous thrombosis, and brain abscess.
- Histology: Necrosis, granulation tissue, skin ulceration.
- Treatment: Debridement, long term IV and oral antibiotics,
- Mortality rates once >50%. Improved with aggressive antimicrobial therapy.

**-THE KEY IS EARLY RECOGNITION.**

#### 2. Rhinocerebral Phycomycosis

- An angioinvasive fungal infection beginning in the sinonasal tract and progressing to intracranial involvement. Caused by *Rhizopus*, *Absidia*, *Mucor*.
- Seen in debilitated patients, diabetics, immunodeficient patients, patients with hematologic malignancies, and renal failure.
- Presents with rhinitis with "sniffles", but progresses over 1-3 days to massively destructive sinonasal lesion with intracranial extension; signs of advancing disease include facial pain, proptosis, ophthalmoplegia, and necrosis of nasal and facial tissue.
- Histology: Broad non-septate hyphae infiltrate soft tissue and vessels; massive necrosis due to thrombosis and ischemia.
- Treatment: Requires early detection, debridement, and implementation of anti-fungals such as amphotericin B.
- High mortality rate.

**-WATCH FOR SNIFFLES IN PATIENTS HOSPITALIZED FOR DIABETIC CRISES.**

#### Lesions Causing Nosebleeds

- \*Pyogenic granuloma (lobular capillary hemangioma)
- \*Juvenile nasopharyngeal angiofibroma
- \*Hemangiopericytoma

**\*Pyogenic granuloma (lobular capillary hemangioma):**

- Benign polypoid vascular proliferation, usually in adults, uncommon in children.
- Anterior nasal septum most common location.
- Uncertain etiology, but associations with trauma and pregnancy.
- Gross: Smooth, lobulated, red polypoid mass, up to 1.5 cm.
- Histology: Arborizing proliferation of central capillaries with tiny branches peripherally.
- Treatment: Local excision. Rarely recurs.

**\*Juvenile Nasopharyngeal Angiofibroma:**

- Benign neoplasm composed of a mixture of vascular and fibrous tissue with locally destructive growth.
- Virtually exclusively in males, usually second decade; rare over age 25. If seen in female, check karyotype.
- Originates in lateral or upper posterior nasopharynx.
- Presents with epistaxis and unilateral nasal obstruction. Less commonly proptosis, anosmia, pain, or facial deformity.
- Histology: Gaping antler-like thin walled vessels, surrounded by collagenous stroma with stellate fibroblasts.
- Treatment: Complete resection if at all possible. Recurrent or progressive tumor may cause destruction of vital structures, especially with intracranial extension. Complete resection, usually with preoperative embolization, is desirable. Estrogen therapy (hormonal modulation) and radiation have been used for recurrences.
- Mortality rate: 5%.

**Hemangiopericytoma:**

- Vascular neoplasm of variable biologic behavior. May be seen anywhere in soft tissue of body. In sinonasal locations it is usually more indolent than in other sites. Propensity for recurrence more than for metastasis.
- Head and neck is a common location (up to 25% of hemangiopericytomas in this area).
- Histology: Staghorn shaped vessels with plump spindle cell background. The histologic features are not very predictive of behavior.
- Treatment: Complete resection; recurrence is common.

**Lesions Causing Nasal Obstruction/ Masses**

Sinonasal Inflammatory Polyps Sinonasal Papillomas Rhabdomyosarcoma Olfactory Neuroblastoma Melanoma
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**Sinonasal Inflammatory Polyps:**

- Non-neoplastic inflammatory swellings of sinonasal mucosa.
- M=F. Seen at any age, but most often in adults over 20 years and rare in children less than 5 years.
- Unilateral or bilateral.
- Symptoms: nasal obstruction, rhinorrhea, headaches.
- Etiologic links: Cystic fibrosis, infections, diabetes, aspirin intolerance.
- Gross: Edematous fleshy polypoid pink gray masses protruding into nasal cavity.
- Histology: Intact surface epithelium (usually respiratory type, covering edematous gelatinous stroma lacking seromucinous glands. The stroma contains inflammatory cells including plasma cells, lymphocytes, and many eosinophils.
- Treatment: Surgical excision. Addressing etiologic factors.

### **Sinonasal Papillomas:**

- Benign neoplasm of the sinonasal mucosa, composed of squamous or columnar cells.
- Although benign these lesions may be locally very destructive and potentially fatal due to encroachment on vital structures.
- Occur over wide age range but very uncommon in children. Usually unilateral but may occasionally be bilateral. More common in males.
- Symptoms: Nasal obstruction, epistaxis, pain.
- Gross: Polypoid masses covered with mucosa. May have a somewhat warty surface. May or may not be visible on direct nasal examination.
- Radiographic: CT and MRI studies important to delineate extent. These may fill the nasal cavity and extend into multiple sinuses. May erode bone by pressure.
- Histology: Three types are recognized, fungiform, inverted, cylindrical cell; all may be lumped together under term "sinonasal papilloma". Excessive proliferation of benign appearing squamous or columnar epithelium. Watch for dysplasia or foci of squamous cell carcinoma in these lesions.
- Pathologists should examine all of the tissue if possible to exclude areas of malignant change.
- Surgeons should completely excise with a margin of normal mucosa to prevent recurrence.
- Incidence of malignant change: <10% for any histologic type. Usually it is focal change and does not alter the surgical approach..

### **Rhabdomyosarcoma:**

- Malignant tumor with skeletal muscle differentiation.
- Most common sarcoma in the pediatric, adolescent, and young adult populations.
- Most common sarcoma of head and neck region (45% of rhabdos occur there).
- M=F. Most common in first and second decades, rare beyond 5<sup>th</sup> decade.
- Sites: Orbit>nasopharynx>ear (middle ear & mastoid)>sinonasal cavity.
- Symptoms vary with location: nasal obstruction and epistaxis are associated with sinonasal.
- Gross: "Sarcoma botryoides" is term given to polypoid tumors occurring in cavities such as sinonasal tract, middle ear, etc. They look very similar to nasal polyps, often like a cluster of grapes. Have white to gray gelatinous appearance. Often necrotic.
- Histology: Often mimic nasal polyps because of gelatinous myxoid appearance, but they are more cellular (mostly small undifferentiated, primitive cells). The key to dx is the rhabdomyoblast which demonstrates skeletal muscle differentiation (cross-striations or antigenic evidence of production of skeletal muscle associated proteins such as actin and myoglobin).
- Treatment: Nonradical surgery +chemotherapy + radiation.
- Outcome: Stage dependent survival: 5 year survivals for Stage I (localized disease with complete resection-83%; Stage IV (distant metastases)-20%.

**-BE SUSPICIOUS OF A "NASAL POLYP" IN A VERY YOUNG CHILD!**

### **Olfactory Neuroblastoma:**

- Malignant neoplasm derived from the olfactory membrane of the sinonasal tract. Demonstrates primitive neuronal differentiation.
- M=F; at any age from infancy to old age; two peaks: second and sixth decades.
- Symptoms: Unilateral nasal obstruction, epistaxis, headaches, visual disturbances.
- Radiologic: May achieve large size with extensive involvement of nasal cavity and sinuses, and some with orbital and intracranial extension. Epicenter of lesion should be cribriform plate/ ethmoid sinuses.
- Gross: Bulky polypoid mass in nasal cavity, often with hemorrhage or necrosis.
- Histology: Primitive small round blue cell tumor with neurofibrillary background. Resembles other types of neuroblastoma.
- Treatment: Radical surgery followed by full course of radiotherapy. Chemotherapy +/-.
- Prognosis related to stage and grade: Mostly locally aggressive, but some metastasize to node, bone, lung, etc. Tumors confined to nasal cavity and resected completely->90% 3-year survival; tumors with extension beyond sinonasal cavities-<50% 3-year survival.

## Melanoma:

- Malignant melanoma occurs as a primary malignancy in the sinonasal tract. It is relatively uncommon, but should always be in the clinical and pathologic differential diagnosis of a nasal mass.
- The prognosis is poor in sinonasal melanoma. The staging criteria and outcomes are not synonymous with those of cutaneous melanoma.

### Midline Destructive Diseases

Wegener's Granulomatosis  
Midline Malignant Reticulosis  
Granulomatous Infections

## Wegener's Granulomatosis:

- Non-neoplastic idiopathic aseptic necrotizing granulomatous disease. Predilection for upper and lower respiratory tracts and genitourinary tract.
- May be systemic or localized.
- M>F (except in laryngeal Wegener's). Most common in 4<sup>th</sup> and 5<sup>th</sup> decades.
- Most common H&N sites are sinonasal.
- Symptoms of sinonasal involvement: Sinusitis, purulent rhinorrhea, obstruction, pain, epistaxis.
- Lab studies: ESR elevated, renal functions elevated if GU involved. Antineutrophilic cytoplasmic antibody elevated.
- Radiologic: Sinus opacification with bone and soft tissue destruction.
- Gross: Ulcerative and crusted lesions, often septal perforation, necrotic tissue, loss of tissue. Saddle nose in advanced cases.
- Histology: Granulomatous inflammation with vasculitis; geographic necrosis.
- Treatment: Limited disease with steroids or cyclophosphamide, good prognosis. Systemic disease requires high dose steroids and cyclophosphamide, poor prognosis.

## 2. Midline Malignant Reticulosis:

- A highly destructive angiocentric form of T-cell lymphoma associated with extreme tissue loss in the sinonasal tract.
- Also previously called lethal midline granuloma.
- M>F. Most common in 5<sup>th</sup> & 6<sup>th</sup> decades.
- Radiologic findings are same as Wegener's.
- Gross: Soft polypoid masses in sinonasal cavity associated with ulceration, purulent and necrotic material. Bone and soft tissue destruction may be extensive if dx delayed.
- Histology: Polymorphous lymphoid infiltrate with vasculitic component and extensive necrosis.
- Often diagnosis delayed due to necrotic biopsies or mistaken for infection or Wegener's.
- Treatment: Localized with radiation; systemic with chemotherapy and radiation.
- Prognosis: Localized: > 50% 5-year survival; systemic: rapidly lethal.

## Infectious Processes:

- Rule out fungal and mycobacterial infections

## Laryngeal Nodules and Polyps:

- Non-neoplastic stromal reactive process related to trauma or inflammation.
- "Nodules" are bilateral, follow chronic voice abuse, and involve ant or middle third of TVC.
- Polyps are single, follow voice abuse, infection, alcohol, smoking, or hypothyroidism, and are most common in the middle third of the TVC, but may occur in ventricle.
- Gross: Shiny fusiform swelling, sessile, or pedunculated lesion, few mm's to several cm's.

- Histology: The changes are largely stromal, with myxomatous degeneration, fibrosis, vascular proliferation, or hyalinization. Overlying epithelium may be thickened or keratotic.
- Treatment: Voice therapy for nodules; surgery for polyps.

### **Laryngeal Papillomatosis:**

- Benign exophytic neoplastic growth of squamous epithelium
- Adult type: Single lesion, rarely recurs or spreads.
- Juvenile type: Multiple lesions with extensive involvement of larynx, trachea, and sometimes bronchial tree. High recurrence rate. May regress with age. HPV associated; acquired at birth.
- Gross: Exophytic warty growths on mucosa. Tan-white and friable.
- Histology: Papillary fronds of squamous epithelium with fibrovascular cores.
- Treatment: Surgery (laser), antiviral therapy.
- Prognosis: Multiple recurrences usual; mortality 2-14%. Risk of malignant transformation: 2%.

### **Contact Ulcer of Larynx:**

- Benign, tumor-like condition, most common on posterior aspect of TVC, one or both.
- M>F. Mostly adults.
- Etiology: Vocal abuse, acid reflux from GI tract, post-intubational reaction.
- Gross: Ulcerated polypoid or nodular mass, beefy-red to white-tan, up to 3 cm in size.
- Histology: Ulcerated lesion with exuberant granulation tissue.
- Treatment: Identify and treat underlying cause.
- Location at posterior TVC clinically helped distinguish from cancer, which is usually anterior.

### **Tumors of Salivary Glands**

Benign Mixed Tumor  
 Warthin's Tumor  
 Mucoepidermoid Carcinoma  
 Adenoid Cystic Carcinoma  
 Acinic Cell Carcinoma

### **Benign Mixed Tumor:**

- Benign neoplasm of heterogenous type, with epithelial and mesenchymal components.
- Most common neoplasm of salivary glands (40-70% of all tumors).
- F>M. most common in 3<sup>rd</sup> to 6<sup>th</sup> decades.
- Parotid tail most common site, but may be seen anywhere salivary tissue is found.
- Presents as slow growing firm mass that may be present for many years. May be very large.
- Gross: Cut surface may be firm, white and cartilaginous; usually 1-2 cm.
- Histology: Ductal structures, sheets of epithelial cells, chondromyxoid matrix. Lacks capsule.
- Treatment: Surgical excision with margin of normal tissue to prevent local recurrence.
- Prognosis: Risk of recurrence. Rarely malignant transformation to carcinoma.

### **Warthin's Tumor:**

- Benign tumor with epithelial and benign lymphoid tissue component.
- 5-6% of all salivary tumors.
- M>F. 5<sup>th</sup> and 6<sup>th</sup> decades most common.
- Almost exclusively in parotid gland. 14% bilateral.
- Presents as soft fluctuant mass.
- Gross: Usually partially cystic, filled with motor oil like fluid; fleshy soft red tan tissue.
- Histology: Oncocytic epithelium in papillary fronds; benign lymphoid tissue.
- Treatment: Surgical excision. Totally benign lesion.

**Mucoepidermoid Carcinoma:**

- Most common malignant salivary tumor.
- M=F: wide age range, including children; most common in 3<sup>rd</sup> to 6<sup>th</sup> decades.
- Most often in the parotid, but anywhere there is salivary tissue.
- High, intermediate, and low grades effect prognosis.
- Gross: Infiltrative but circumscribed white-tan mass, often partially cystic, with mucus.
- Histology: Epidermoid cells and mucus cells.
- Treatment: Surgery!
- Prognosis: Low and intermediate grade usually cured with resection; may recur but mets uncommon; high grade aggressive, with mets common- 40% 5 year survival.

**Adenoid Cystic Carcinoma:**

- Most common malignant tumor of submandibular gland. Most are found in parotid or SMG.
- Seen wherever there is salivary gland tissue.
- Extremely difficult to treat adequately because perineural invasion is usually present. Pain is a clue to diagnosis.
- Gross: Irregular firm mass. Not very distinctive.
- Histology: Classic pattern is cribriform, or swiss cheese like. Holes of swiss cheese filled with basement membrane material.
- Treatment: Aggressive surgery; radiation often used.
- Prognosis: Poor in essentially all cases. Patients may live for 10 years but virtually all recur and eventually die from disease. Metastases occur late in disease; death usually within 1 year of mets.
- NO ONE IS CURED OF ADENOID CYSTIC CARCINOMA.

**Acinic Cell Carcinoma:**

- Usually a low grade malignant tumor, less common than the above lesions.
- (0% occur in parotid.
- May be seen at any age, including pediatric. F>M. Most in 4<sup>th</sup>/ 5<sup>th</sup> decades.
- Presents as painless or painful mass.
- Gross: Well-circumscribed or encapsulated yellow-tan mass, often with cystic areas.
- Histology: Cells are similar to the acinar serous cells of the normal gland, and have zymogen granules which give the cytoplasm a purple granular appearance on H&E.
- Treatment: Surgery.
- Prognosis: Indolent tumors, usually cured by surgery. 6% mortality rate.