Vascular Tumors of the Head and Neck

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Introduction

- Many different types of neoplasms
- Share common etiology with vascular system
- Benign, malignant, others
Benign Tumors

- Vascular Birthmarks
  - Hemangiomas
  - Vascular Malformations
- Nasopharyngeal angiofibromas
Vascular Birthmarks

- History
Vascular Birthmarks

Classification system

- Hemangioma vs. malformation
- Based on clinical, cellular, biologic factors
- Older terms – “capillary”, “juvenile”, “strawberry”, “cavernous”
Vascular Birthmarks

- Classification system
  - Superficial vs. Deep Hemangiomas

<table>
<thead>
<tr>
<th>Hemangioma</th>
<th>Old Terminology</th>
<th>Malformation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Capillary</td>
<td></td>
<td></td>
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<tr>
<td>Strawberry</td>
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<td></td>
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<tr>
<td>Port-wine</td>
<td></td>
<td>CAPILLARY</td>
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<td>Capillary-cavernous</td>
<td></td>
<td></td>
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<td></td>
<td>VENOUS</td>
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<td>Hemangio-lymphangioma</td>
<td></td>
<td>LYMPHATIC</td>
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<td>Lymphangioma</td>
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<td>Arteriovenous</td>
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Hemangiomas

- Most common tumor of infancy (10%)
- Slight female predominance
- 60% arise in head and neck – cosmetic concerns
Hemangiomas

- Clinical presentation for diagnosis
  - Not seen at birth
  - Precursor lesion
  - Proliferative phase
  - Involution phase
  - Superficial vs. deep
Complications from Hemangiomas

- Occur in 20%
  - Ulceration
  - Compression of vital structures
  - High-output cardiac failure
  - Bleeding
    - Kasabach-Merritt syndrome
Laryngeal Hemangiomas

- Usually in the subglottis
- Healthy infant with biphasic stridor (croup)
- Behave similarly
- 50% with cutaneous counterpart
Diagnosis of Hemangiomas

- History and physical examination
- Certain cases – radiology
  - Ultrasound, MRI
  - Large facial hemangiomas – Dandy-Walker
Treatment of Hemangiomas

- Why and when to treat?
  - Normal skin in 50% that involute within 5 years
  - Other 50%-- 80% substantial deformity

- Pro’s and Con’s
Treatment of Hemangiomas

- Observation
  - Serial photography important to document involution
  - Regular visits with reassurance
Treatment of Hemangiomas

- Systemic steroids
  - Careful selection criteria
  - Prednisone 2-4mg/kg for up to 6 weeks
  - Varied results (30%)
  - Side effects
Treatment of Hemangiomas

- **Intralesional steroids**
  - Usually for vision threatening lesions
  - Combination of beta-methasone and triamcinolone
Treatment of Hemangiomas

- Pulse-dye lasers
  - Useful for superficial variety
  - Good for ulcerations/residual cosmesis
Treatment for Hemangiomas

- **Surgery**
  - Eyelid lesions, bulky lesions, vermilion border, nasal tip, eyebrow
  - CO2 laser for subglottis
Treatment of Hemangiomas

- Arterial embolization
- Radiation therapy
- Alpha-2b interferon
Vascular malformations

- Capillary, venous, arterial, lymphatic, mixed
- By definition—present at birth
- No proliferative or involution phase
- Commensurate growth
Capillary malformations

- Older term—“port-wine stain”
  - Usually in trigeminal distribution
  - Most isolated anomalies
  - Sturge-Weber syndrome
Treatment of Capillary Malformations

- Cosmetic concealing makeup
- Tattooing
- Surgical excision (tissue expanders)
- Pulse dye-laser
Venous Malformations

- Diagnosis is clinical – palpation
- Treatment dependent on location (surgery and sclerotherapy)
Lymphatic malformations

- Older terms—“cystic hygroma”, “lymphangioma”
- Can expand with URI
- Surgical treatment is mainstay
  - Picibanil (OK-432)
Arteriovenous malformations

- Usually clinically apparent
- Embolization and surgical resection
Nasopharyngeal Angiofibroma

- Most common benign tumor of nasopharynx
- Older term—"juvenile nasopharyngeal angiofibroma", "JNA"
- Presentation: recurrent epistaxis/nasal congestion, hearing loss, orbital, CN
- Arise where sphenoidal process of palatine bone meets horizontal ala of vomer
Nasopharyngeal Angiofibroma

- Diagnosis is made by clinical and radiographic findings
  - CT/MRI
  - Biopsy - rarely indicated
  - Angiography
Nasopharyngeal Angiofibroma

- Angiography

Histology
Nasopharyngeal Angiofibroma

- **Treatment**
  - Embolization and surgery
    - Autologous blood/Cell Saver
  - Approaches
    - Transnasal endoscopic, lateral rhinotomy/MFD with medial maxillectomy or LeFort I, transpalatal, facial translocation/maxillary swing, infratemporal approaches, craniotomy
  - Radiation therapy
  - Chemotherapy
Malignant Vascular Tumors

- Angiosarcoma
  - Extremely rare (50% in head and neck)
  - Prognosis on tumor size, grade, margins
  - Radiation minimally effective
  - Sinonasal tract less aggressive
  - Poor survival
Malignant Vascular Tumors

- Hemangiopericytoma
  - Pericytes of Zimmerman
  - 25% in head and neck
  - Surgical treatment
  - Grade important on prognosis
  - Radiation/chemotherapy for selected cases
Malignant Vascular Tumors

- Kaposi’s Sarcoma
  - Viral-induced
  - Four entities
    - Classic
    - Endemic
    - Immunosuppressed
    - AIDS-related
  - Surgery, chemo, radiation, sclerotherapy
Paragangliomas

- Named for anatomic location
- Arise in paraganglionic tissue (neural crest)
- Type I cells (chief) – APUD cells – catecholamines
- Type II cells (sustentacular)
- Clusters together– “Zellballen”
- Malignancy is clinical
Carotid paragangliomas

- Most common of head and neck (60%)
- Multicentric 10%, malignant 10%
- Familial (AD) 20% -- more multicentric
- Painless mass at SCM, immobile superior-inferior direction
- May produce catecholamines
Carotid paragangliomas
Carotid paragangliomas

- Treatment
  - Surgery
    - Mortality 8%, >5cm tumors had more complications
    - Preop workup key – vascular surgeon, anesthesia
    - Embolization – controversial
  - Radiation
Vagal paragangliomas

- Most commonly at nodose ganglion
- Painless mass at angle of mandible present for many years – enlarging may get Horner’s, CN XII, hoarseness
- More multicentric (25%)
- Malignancy (18%)
- None produce catecholamines
Vagal paragangliomas
Laryngeal paragangliomas

- Usually from superior laryngeal paraganglia from aryepiglottic fold
- Hoarseness and dysphagia common
- High rates of malignancy
- Wide local excision or partial laryngectomy