Granulomatous Diseases of the Head and Neck

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Granulomatous Inflammation

- A type of chronic inflammation characterized by concentric layers of cells consisting of specialized macrophages called epithelioid cells and multinucleated giant cells surrounded by lymphocytes and fibroblasts; however, the inflammatory process can be more diffuse without discrete classic granuloma formation.

- Stimulus can be foreign body or persistent microorganism which evades destruction; multiple etiologies exist.
Outline

- Infectious
  - Bacterial
  - Fungal
  - Parasitic
- Trauma/Foreign Body
- Neoplastic
- Inflammatory Disease of Unknown Etiology
- Autoimmune/Vasculitic Disease
Cat Scratch Disease and Bacillary Angiomatosis

- *Bartonella henselae* is the most common organism
- CSD usually self-limited in children and young adults requiring no specific treatment
- BA occurs in immunocompromised patients and requires antibiotic treatment; visceral involvement termed peliosis hepaticus
Rhinoscleroma

- **Organism:** *Klebsiella rhinoscleromatis*
- Three stages: catarrhal, granulomatous, sclerotic
- Treatment is with tetracycline or cipro
- Nasal involvement universal; paranasal sinus involvement much less common
Leprosy (Hansen’s Disease)

- Organism: *Mycobacterium leprae*
- Patterns of manifestation include lepromatous, tuberculoid and mixed
- Rare in United States
- Treatment is with 2-5 years of antibiotics
Nontuberculous Mycobacteria

- Pathogens include M. scrofulaceum, M. avium complex, M. kansasii, M. marinum
- Many manifestations; commonly lymphadenitis in children or severe systemic infection in immunocompromised
Tuberculosis

- Extrapulmonary tuberculosis most commonly affects infants, children and immunocompromised.
- Most common extrapulmonary manifestation is scrofula.
Actinomycosis

- Cervicofacial disease most commonly secondary to dental infection, intraoral trauma or manipulation
- Characteristic sulfur granules; classically perimandibular soft tissue infection or abscess with draining sinus and possible bone involvement
- Requires anaerobic processing of culture specimens
- Can be treated with ampicillin, Pen G or Clindamycin
Syphilis

- Organism: spirochete *Treponema pallidum*
- Three stages: primary, secondary, tertiary
- Transmitted sexually or transplacentally
- Treatment is with penicillin
Histoplasmosis

- Organism is *Histoplasma capsulatum*
- Risk factor is exposure to soil enriched with bat or bird excrement
- Manifestation of infection depends on number of organisms inhaled and immune status
- May cause mediastinal granulomatosiis and fibrosing mediastinitis
- Treatment is with Amphotericin B or Itraconazole depending on severity of disease
Candidiasis

- Many different Candida species can be pathogenic
- Can cause oral thrush or candida esophagitis
- Candida may also be part of normal flora of oral cavity
- Treatment is usually via topical therapy; systemic therapy may be required with severe infection or immunocompromised patients
Blastomycosis

- *Blastomyces dermatitidis*: dimorphic fungus found in moist soil
- Extrapulmonary cutaneous disease usually occurs in conjunction with pulmonary disease
- May involve multiple organ systems especially in immunocompromised patients
Coccidiomycosis

- Coccidiodes immitis is a fungus that lives in dry, desert soil
- Infection is via inhalation of arthrospores
- Can be asymptomatic or lead to pulmonary infection which can be severe
- May manifest as skin lesion in which case pulmonary or CNS involvement should be suspected
- Treatment is via Amphotericin B or one of the azoles
Phycomycosis

- Sinonasal mucormycosis with etiologies including *Aspergillus* sp. or organisms from the family Mucorales: *Rhizopus, Rhizomucor, Mucor, Absidia* and *Cunninghamamella*
- Susceptible patients are immunocompromised especially patients with DKA
- Spectrum of disease ranging from rapidly progressive and fatal to indolent invasive course
- Symptoms include facial anesthesia, headache, ophthalmoplegia, facial necrosis and obtundation
- Physical exam may reveal white, insensate nasal mucosa, black necrotic mucosa; changes prominent on middle turbinate, hard palate
- Treatment is with amphotericin B and urgent and aggressive debridement
Leishmaniasis

- The bite of a sandfly carrying various leishmania species is the etiology of leishmaniasis
- Visceral (kala-azar), cutaneous, and mucocutaneous (Espundia)
- Treatment is quite toxic
Myiasis

- Infestation with maggots of screw worm or bot fly
- Furuncular, creeping dermal myiasis
- Treatment consists of surgical debridement
Infectious Etiology

- **Bacterial**
  - Cat Scratch Disease
  - Rhinoscleroma
  - Leprosy
  - Nontuberculous Mycobacteria
  - Tuberculosis
  - Actinomycosis
  - Syphilis

- **Fungal**
  - Histoplasmosis
  - Candidiasis
  - Blastomycosis
  - Coccidiomycosis
  - Phycomycosis

- **Parasitic**
  - Leishmaniasis
  - Myiasis
Intubation Granuloma

- Almost universally involves vocal process of arytenoid
- Hoarseness and foreign body sensation common
- Voice rest, control of irritant exposure, possible surgical excision
Teflon Granuloma

- 2-3% of patients receiving Teflon for unilateral vocal cord paralysis develop increasing dysphonia or even airway obstruction secondary to hyperintense granulomatous response to Teflon
- Treatment is via endoscopic removal, laser vaporization or open surgical removal
Trauma/Foreign Body

- Intubation Granuloma
- Teflon Granuloma
Langerhan’s Cell Histiocytosis

- Formerly included:
  - Eosinophilic granuloma (usually monostotic osteolytic lesion with predilection for skull)
  - Hand-Schuller-Christian disease (classic triad of multiple bone lesions, exophthalmos and diabetes insipidus)
  - Letterer-Siwe disease (acute, disseminated)
Lobular Capillary Hemangioma (Pyogenic Granuloma)

- Neither infectious or granulomatous
- Exact etiology unknown
- Solitary glistening red papule prone to bleeding and ulceration
- Occurs most often on trunk, head, neck, upper extremities
- Oral cavity lesion may develop in pregnancy ("pregnancy tumor")
Necrotizing Sialometaplasia

- Non-neoplastic condition of the salivary gland, typically the minor salivary glands of the palate
- Is self-limiting
- Etiology unknown but may be related to trauma or vomiting
- Typically an ulcerative lesion on the hard palate
- Can be confused with squamous cell carcinoma and mucoepidermoid carcinoma on histopathologic evaluation
Neoplastic

- Langerhan’s Cell Histiocytosis
- Lobular Capillary Hemangioma
- Necrotizing Sialometaplasia
Sarcoidosis

- In the United States, the disease is more prevalent in African Americans; slight female preponderance
- Disease manifestations and course variable; pulmonary involvement almost universal
- Lofgren’s syndrome: hilar lymphadenopathy and erythema nodosum; Heerfordt’s syndrome (uveoparotid fever): anterior uveitis/parotid swelling/facial nerve palsy/fever
- Otolaryngologic manifestations occur in 10-15% of patients—cervical adenopathy, parotid swelling and facial nerve palsy are the most common findings
- High rate of spontaneous remission
- Diagnosis via BAL with high CD4/CD8 ratio and transbronchial lung biopsy
- Elevated ACE suggestive but may not be present
- Treatment via steroids/methotrexate
Idiopathic Midline Destructive Disease

- Rare spectrum of lymphoproliferative disorders causing destruction of the nose, paranasal sinuses, palate and facial soft tissue
- Controversy exists over whether all cases of IDMM are lymphoma or Wegener’s
- Treatment is with XRT
Inflammatory Diseases of Unknown Etiology

- Sarcoidosis
- Idiopathic Midline Destructive Disease
Wegener's Granulomatosis

- Classically the triad of pulmonary, renal, and head and neck manifestations
- Nose and paranasal sinuses most commonly affected site in the head and neck
- Diffuse crusting of the nose and nasopharynx; removal of crusts leaves friable mucosa
- Orbital involvement common: nasolacrimal duct obstruction/proptosis due to pseudotumor
- Subglottic involvement apparent in 1/5 of patients
- Diagnosis involves serum ANCA testing and biopsy (remove all crusts and biopsy every turbinate)
- Treatment is with steroids and cyclophosphamide with long term bactrim
Relapsing Polychondritis

- Patients with this rare disorder produce antibodies to type II collagen
- Ear exam in acute case may reveal red, swollen and tender ear with sparing of the lobule; over time, the ear becomes droopy
- Nasal chondritis may lead to saddle nose deformity
- 50% of patients have laryngotracheal disease with potential dynamic collapse of the trachea
Sjogren’s Syndrome

- Affects salivary and lacrimal glands
- May be primary (sicca syndrome) or secondary
  - (in association with other autoimmune diseases)
- May evolve into lymphoma
- 90% patients women
- Monitor for signs of lymphoma
- Check for other autoimmune diseases
- SS-A, SS-B antibodies
Autoimmune/Vasculitic Disorders

- Wegener’s Granulomatosis
- Relapsing Polychondritis
- Sjogren’s Syndrome
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