Granulomatous Diseases of the Head & Neck

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Grand Rounds Presentation
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History

- From Latin “granulum” - small particle, grain
- Hippocrates - describes syphilis
- Tuberculosis found in Egyptian Mummies
- Margret - 17 cent. - coins term “miliary”
- Koch - stains and describes granuloma
- Bergey - Actinomyces isolated in vivo and sulfur granules described
- Etiology of many diseases continue to evolve today
Pathophysiology

- Neutrophils remove foreign bodies
- Indigestible bodies, prolonged inflammation solved by granulomatous inflammation
- Principal cells - Macrophages, Lymphocytes
- Phagocytosis, loss of antigenicity, then epitheloid change
- Fusion to Multinucleated giant cells
- Langhan’s giant cell, foreign body giant cell
- Lymphocytes surround giant cells, other inflammatory cells may be associated
Pathology - granuloma
Pathology - giant cells
Presentation, workup

- Present as nonhealing ulcer or mass
- FNA shows granulomatous inflammation
- Hx: Fever, night sweats, weight loss, anorexia, arthralgia, malaise
- Foreign travel, immunosuppression risks
- PE: LAD-axillary, inguinal, cervical
- Labs: CBC, ESR, ANA, UA, others
- CXR, excisional bx if needed for cxs, dx
- Rheumatology, ID consults early
- Tx vastly differs, exact dx necessary
Fungus - Hisplasma capsulatum

- Endemic to U.S.- Mississippi, Ohio River V.
- Inhalation of spores, usually no disease
- Viral type sx acutely, then may go into chronic phase with constitutional sx
- Disseminated disease evident on CXR
- Half of adults with H&N manifestations
- ENT - dysphagia, sore throat, hoarseness, gingival pain,
- Lips, gingiva, tongue, pharynx, larynx
- Ulcer with “heaped up” margins, verrucoid
- Swab or bx and cx, tx with amphotericin B
Fungus - Blastomyocosis dermatidis

- SE, central, mid-atlantic US, <<histoplasma
- Middle aged males, usually asx
- Chronic disease - GU, lung, bone, skin
- Skins lesions verrucoid, with scarring
- ENT mucosal involvement very rare, Larynx/pharynx w/ erythematous hyperplasia, TVC fibrosis and PC fistula late stage
- CXR abnormal 75% with nodular infiltrates
- Dx with sputum culture, skin scraping - Tx with Ampho B
Fungus-Phyco/Mucormycosis

- Immunocompromised patients
- Mucor, Rhizopus, Absidia sp; ubiquitous
- Begins in sinus, locoregional spread
- Sx: Face pain, bloody rhinorrhea, fever, edema
- Diplopia, obtundation, death
- CN neuropathy @ 90% mortality
- PE: Face pain, edema, proptosis, poor EOM, nasal mucosa with black eschar
- Dx: bedside emergent mucosa biopsy for staining.....invasion noted on micro
- Tx: CT then OR then ampho B if still alive
Fungus - Aspergillus

- Invasive, subacute, chronic, allergic
- Noninvasive: single sinus w/ dark, thick nasal d/c and fullness as primary complaints
- May progress to granulomatous lesion
- Invasive: Similar to phycomycosis, slower
- CT: inhomogenous sinus density in all forms, calcifications in all but acute form
- Tx: surgical excision, if invasive then emergently with postop amphoB
Fungal - Candida albicans

- Immunocompromised
- Sx: severe odynophagia, dysphagia, laryngitis, angular cheilitis
- Typically white pseudomembrane in oral cavity or oral pharynx
- Dx: swab and micro with culture
- Tx: with nystatin, systemic antifungals for persistent or invasive disease
Candidiasis
Fungus - Rhinosporidium seeberi

- Causative organism of rhinosporidiosis
- Prominent in Southern India/Sri Lanka
- Mucus membranes of nose, palate, conjunct.
- Sx: Chronic rhinitis w/ mucoid d/c, epistaxis
- Polypoid, painless, friable lesion
- Tx: Surgical excision
Rhinocporidiosis
Parasites - Leishmaniasis

- Cutaneous, mucocutaneous involve H&N
- Vector - sandfly
- Cutaneous - papules, then ulcers, resolution usually within 6 months
- Ear/ nose may become chronic destructive
- Mucocutaneous - Central/South America, extremity bite w/ hematogenous spread to NP and OC, necrosis over months to years
- Dx: Biopsy and stain…”Donovan bodies”
- Tx: IV Pentostam x 20 - 30 days
Donovan bodies
Cutaneous Leishmaniasis
Parasite - Myiasis

- Infection w/ maggots of the screw worm fly
- Fly lays eggs in wound, or is inhaled
- In U.S., furuncular form is most common
- Pruritic furuncle develops where eggs laid
- This becomes a nonhealing papule, from which larvae emerge when hatched
- May also occur in nasopharynx, usually reserved for Asian countries
- Dx: microscopic exam
- Tx: Excision and curettage
Parasite - Toxoplasmosis

- Caused by ingestion of *T. gondii* via cat feces, or rare lamb/ pork
- Most patients mount an adequate defense
- May attack any organ system, esp CNS in HIV patients w/ intracranial calcifications
- ENT - Persistent neck mass
- Dx: Biopsy
- Tx: Pyremethemine, Trisulfapyridines
Bacteria - M. tuberculosis

- Spread person to person w/ inhaled droplets
- Most persons...clearance of bug w/o sx
- May form calcified granuloma
- Ghon complex - Ca+ granuloma with hilar LN
- 5% unable to contain bug - active disease
- Pulmonary component dominant
- Cervical LAD MC ENT, B post triangle
- Larynx-1%: arytenoid>TVC>epiglottis>FVC; Sx: cough, hoarseness, weak voice; PE: lesion edematous, ulcerative, or polypoid
Laryngeal Tuberculosis
Bacteria - M. Tuberculosis

- Oral cavity 0.5 - 1.5% : lesions extremely variable, tongue MC site in oral cavity
- Bilateral parotid enlargement common
- Otologic rare: Multiple TM perfs, watery otorrhea, poss mastoiditis
- Dx: Hx, PPD, CXR, sputum stain
- Exc bx of lymph node may be necessary
- Tx: multiple agents for 9-12 months
Tuberculosis - oral cavity
Tuberculosis - otologic
Bacteria - Non TB Mycobacteria

- Kansasii, gordonii, MAI, fortuitum, etc.
- Transmission - soil to mouth/eye
- Usually children, HIV patients
- Children - Corneal ulceration >> scrofula
- LAD unilateral in submandib, preauricular
- May suppurate and/ or fistulize
- Dx by excisional bx with AFB stain/ cx
- Tx: combination rx therapy empirically until cultures back. May excise or curettage, but risk fistula formation
Mycobacterium - scrofula, AFB stain
Bacteria - *M. leprae*

- Leprosy (Hansen’s disease); tropical climates
- Vector - human via nasal secretions, open sores, breast milk
- Tuberculoid form - Widespread peripheral nerve involvement w/ pain, muscle atrophy
- Lepromatous form - cutaneous with hypopigmented concave macules
- Sx: nasal congestion, epistaxis, hoarseness due to mucosal nodules - cartilage collapse, saddle deformity, leonine facies
- Dx - bx : Tx - Dapsone, resistance prominent
Leprosy
Bacteria - Cat Scratch Disease

- *R. henselae, Afipia felis*
- 90% < 18 y/o
- Vesicle or papule w/ regional LAD
- Dx: exposure, primary inoculation site, hist. of biopsy (necr. gran., stellate abscesses)
- Resolves 1-2 months, may need surgery
- Bacillary angiomatosis - same bug, young adults, mostly HIV+, fatal if untreated
- Cutaneous papules or subQ nodules
- Both respond to emycins, doxycycline, rifampin
Bacterial - Actinomycoses

- Aspiration into lung or mucosal contact
- ENT - red, nontender SQ mass, level I
- Over 1/2 pts w/ multiple draining sinuses
- 3/4 w/ constitutional sxs
- Dx: sulfur granules on micro from bx, with characteristic bacterial growth pattern
- Tx: oral pcn, or tetracycline x 2-4 months
- May need surgery to expedite recovery
Actinomycosis, path and lesion
Bacterial - Rhinoscleroma

- *Klebsiella rhinoscleromatis*
- Central America and Eastern Europe
- Prolonged purulent rhinorrhea followed by granulomas in the upper airway which coalesce and lead to sclerosis of the nose, larynx, and tracheobronchial tree
- Dx: bx showing bug in vacuolated histiocytes - Mikulicz cells
- Tx: Streptomycin or tetracycline
- May need dilation procedures
Rhinoscleroma path/lesions
Bacterial - Syphilis

- *T. pallidum*, increased incidence
- Primary - painless chancre lips, tonsil, tongue
- Secondary - disseminated mucocutaneous white macules/papules, acute rhinitis, laryngopharyngitis, OM, alopecia
- Tertiary - gumma as erosive granulomatous lesion.....nasoseptal perf, saddle deform, hard palate perf, laryngeal nodules and ulcers, temporal bone - devascularized - sudden B fluctuating SNHL, vertigo
- Congenital - Hutchinsons incisors, mulberry molars, MR, SNHL, saddle nose deformity
- Dx - Darkfield microscopy, VDRL, FTA - ABS
- Tx: penicillin or tetracycline
Syphilis slides
Traumatic Etiologies

- **Post-intubation granuloma** - adult females, vocal process of arytenoid, hoarse, pedunculated lesion; Tx voice rest v. surgery

- **Pyogenic granuloma** - not a true granuloma
  Bacterial infection after trauma
  Painless friable gingival mass
  Surgical excision for bx or if symptomatic

- **Reparative granuloma** - ? Etiology
  Peripheral - pedunculated submucosal mass
  Central - endosteal, ant to first molar
  Tx - curettage
Post intubation granuloma
Pyogenic granuloma
Foreign Bodies

- **Gout** - urate crystals deposit in soft tissues
  - Tophi in helix/ antihelix, may extrude
  - Polarized microscopy - urate crystals
  - Arthritis may involve cricoarytenoid joints causing throat pain, hoarseness, dysphonia
  - Tx - Colchicine, indomethacin, allopurinol

- **Cholesterol granuloma** - temporal bone/sinus
  - Lack of aeration = cell breakdown = cholesterol deposition and granuloma formation
  - T- bone - asx, CN V - VIII if CPA, cholesteatoma
  - Sinus - congestion, rhinorrhea, facial pain, CT shows smooth walled mass
  - Tx - surgical draining and aeration of site
Gout
Cholesterol granuloma
Cholesterol granuloma v. Cholesteatoma on MRI
Necrotizing Sialometaplasia

- Found anywhere there is salivary tissue
- MC at junction of hard and soft palate
- Sharply demarcated ulcer
- Pathology - Metaplastic epithelial cells lining salivary ducts with *preservation of ductal architecture*
- May be confused with SCCA or mucoep
Necrotizing Sialometaplasia lesion and path
Sarcoidosis

- Black>white, 25/100,000, 30 -50 y/o, F>M
- Involved tissue distorted with noncaseating granulomas causing sx
- Lung, LN, skin, eye MC structures
- Dyspnea, dry cough...90% abnl CXR
- LAD.. Intrathoracic>> cervical
- Skin.. Erythema nodosum, plaques, SQ nodules, lupus pernio
- Bilateral parotid enlargement in 10 %
- 5% w/ supraglottic laryngeal nodules, edema
- Labs: hypergammaglobulinemia, elevated Ca+, LFT, ESR, and/or ACE
- Tx: prednisone or other immunosuppressive rx
Sarcoid slide
Sarcoid slide
Case presentation

- 75 y/o male presents to your office complaining of his left ear being “plugged up”. While listening to him, you notice that he is somewhat dyspneic. He admits that he has felt short of breath for quite some time.
- He has noticed progressive hearing loss over the past 20 years, but denies other otologic complaints. He denies hemoptysis, dysphagia, and pain.
- PMH: COPD, CAD, HTN, Pneumonia
- Allergies: Penicillin, Aspirin
- PSH: Herniorrhaphy - 1954; Cardiac cath - ‘97 “Benign skin cancer” right ear with reconstruction x 3 due to multiple wound dehiscences -1999
- Meds: Captopril, Verapamil, Inhalers
Case Presentation

Left Ear

Right ear
Case Presentation
Case Presentation
Case Presentation

Labs - within normal ranges
Case Presentation

- Assessment - TVC polypoid lesion
- Plan - DL with excision of lesion
- Path - acute and chronic granulomatous inflammation
- Dx - Post intubation granuloma
- Patient’s sx resolved,