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# **EAR AND NOSE INVOLVEMENT IN SYSTEMIC DISEASES**

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# **SYSTEMIC DISEASES**

## **EAR AND NOSE**

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- **Granulomatous processes**
  - Langerhan's cells Histiocytosis
  - Tuberculosis
  - Wegener's granulomatosis
  - Sarcoidosis
  - Churg-Strauss Syndrome
- **Collagen vascular and autoimmune processes**
  - Polyarteritis nodosa
  - Systemic lupus erythematosus
  - Rheumatoid arthritis
  - Sjögren's syndrome

# Wegener's Granulomatosis

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- **systemic and necrotizing granulomatous vasculitis with striking affinity to affect the upper and lower respiratory tracts**
- **histology: necrotizing vasculitis involving small vessels associated with pronounced geographic necrosis and mixed inflammatory cell infiltrate**



# Wegener's Granulomatosis

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- **Lung** 60-90%
- **Kidney** 60-85%
- **Upper respiratory tract** >85%
- **Skin** 40-50%
- **CNS or PNS** 20-30%
- **Eye** 20-40%

# Wegener's Granulomatosis

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- **Nasal involvement (60-80%)**
  - persistent rhinorrhea
  - nasal crusting
  - pain
  - epistaxis
  - congestion
  - septal perforation
  - nasal ulcers

# Wegener's Granulomatosis

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- **Otologic manifestations (19-61%)**
  - otalgia
  - otitis media
  - tinnitus
  - hearing loss ———→
    - vasculitis of cochlear artery
    - perforation of tympanic membr.
    - mastoiditis

**Murty; Clin Otolaryngol, 1990**



# Wegener's Granulomatosis

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- A biopsy should be taken of any suspicious lesions even if an histologic confirmation is difficult
- 126 biopsies in 70 pts.
  - vasculitis+necrosis+granumatosus infl. 16%
  - vasculitis+necrosis 23%
  - vasculitis+ granumatosus infl. 21%

**Devaney et al.; Am J Surg Pathol, 1990**

# Churg-Strauss Syndrome

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- **Eosinophil-rich, granulomatous inflammation and necrotizing vasculitis of the upper respiratory tract affecting small to medium vessels associated with asthma and eosinophilia**
- **3 phases:**
  - **prodromal phase (allergic disease)**
  - **peripheral blood and tissue eosinophilia, chronic eosinophilic pneumonia, eosinophilic gastroenteritis**
  - **systemic vasculitis**



# Churg-Strauss Syndrome

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- **Nasopharyngeal symptoms and signs**
  - **nasal polyps without diffuse mucosal destruction (as Wegener's granulomatosis)**
  - **persistent rhinorrhea**
  - **pain**
  - **congestion**
  - **fibrinoid, necrotizing epithelioid eosinophilic granuloma**

# Sarcoidosis

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- **Chronic granulomatous disorder (non-necrotizing) of unknown etiology that may involve any organ system**
- **The spectrum of otolaryngoiatric lesions is extraordinary broad**
- **Ear and nose involvement > 33%**

# Sarcoidosis

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- **Nasopharyngeal symptoms and signs (10-20%)**
  - persistent rhinitis
  - sinus congestion  
(unresponsive to inhaled decongestant and topic steroids)
  - crusting of nasal mucosa
  - septal perforation
  - nasal mucosal nodules or papules
  - polypoid masses
  - *lupus pernio*