

Sarcoidosis

- **Otologic manifestations**
 - **bilateral parotid gland enlargement**
 - **sicca syndrome**
 - **sinusitis**
 - **cervical lymphadenopathy**
 - **cranial nerve palsies**
 - ***lupus pernio***

Langerhan's Cells Histiocytosis

- **Group of disorders of the reticuloendothelial system that are characterized by a proliferation of cytologically benign histiocytes**
 - **Eosinophylic granuloma**
 - **Hand-Schüller-Christian disease**
 - **Letterer-Siwe disease**
- **Unknown etiology (immunological basis?)**

Langerhan's Cells Histiocytosis

- Otologic manifestations occur in 15-61%
 - mastoid (common site)
 - otic capsule
 - facial nerve
- It could be the initial presentation of the disease
- Diagnosis suggested by an inflammatory disorder of the middle ear and mastoid that does not respond to antibiotic therapy

Cunningham MJ et al.; Arch Otolaryngol Head Neck Surg 1989

Langerhan's Cells Histiocytosis

- **Otologic symptoms and signs**

- otorrhea
- postauricular swelling
- hearing loss
- vertigo
- granulation tissue or aural polyps in the external auditory
- perforation of the tympanic membrane
- otitis media or externa
- mastoid fistula

Langerhan's Cells Histiocytosis

- Rx: destructive lesions in the mastoid and temporal bones
- Because the surface of the granulation tissue often shows injection, necrosis and fibrosis, biopsy specimens should be taken from deeper part of the lesion



Histiocytes

Eosinophils, Plasma Cell, PMN, Multinucleated Cells

Systemic lupus erythematosus (LE)

- **Widespread fibrinoid degeneration of the collagen fibers that affects the mesenchymal tissues**

Classification

- **Chronic cutaneous LE**
 - **discoid (DLE)**
 - **generalized DLE**
 - **hypertrophicD LE**
- **Subacute cutaneous LE**
- **Acute cutaneous LE**

Systemic lupus erythematosus

- Typically involves the bridge of the nose, malar areas, ears and scalp (15%)
- Otologic manifestations
 - chronic otitis media
 - hearing loss
 - disequilibrium

Systemic lupus erythematosus

- **Contributing factors**
 - ultraviolet light
 - cold
 - thermal burn
 - physical trauma
- **Lesion: well-circumscribed papule or plaque with follicular plugging**

Rheumatoid arthritis

- **Chronic systemic inflammatory disease that affects the small joints in a symmetric fashion**
- **Extrarticular manifestation:**
 - vasculitis
 - muscle atrophy
 - subcutaneous nodules
 - lymphadenopathy
 - splenomegaly
 - leukopenia

Rheumatoid arthritis

- **Otologic manifestation (rare)**
 - loosening of the transducer mechanism
 - vestibuloauditory dysfunction
- **Relationship between Rheumatoid Arthritis and inner-ear dysfunction is not clear**

Conclusions

- **The incidence of ear and nose involvement in systemic diseases varies between the diseases**
- **Because of the difficult diagnosis, a biopsy specimen is always needed**
- **The treatment does not vary from the generalized forms, except in rare cases (mastoid surgery for Hx)**