Sarcoidosis
Dr. Salah Ahmed

- a multisystem granulomatous disease of unknown etiology characterized by noncaseating granulomas in many tissues and organs

Epidemiology:

- it occurs throughout the world
- affects both sexes and all races and ages
- common in adults younger than 40 years of age
- one of the pulmonary disorders that common among nonsmokers

Etiology and Pathogenesis:

- etiology remains unknown
- three factors involved in pathogenesis (suggested)

1- immunologic abnormalities: suggested because

- high level of CD4+ helper T cells that secrete cytokines such as IL-2, IFN-γ

2- genetic predisposition: suggested because

- familial and racial clustering of cases
- association with certain human leukocyte antigen (HLA) genotypes (e.g., class I HLA-A1 and HLA-B8)

3- exposure to certain environmental agents:

- several "antigens" have been proposed as the inciting agent for sarcoidosis (e.g., viruses, mycobacteria, Borrelia, pollen), but no evidence

Morphology:

- noncaseating epithelioid granuloma:
collection of epithelioid cells rimmed by
an outer zone of CD4+ T cells. also
multinucleated giant cell, fibroblasts
- Two other microscopic features are sometimes seen in the granulomas:
(1) Schaumann bodies, laminated concretions composed of calcium and proteins
(2) asteroid bodies, stellate inclusions enclosed within giant cells

Organs involvement:

Lungs	Upper airways	Lymph nodes
Skin		
Eyes	Liver	Spleen
Bones		
Salivary gland	Heart	Nervous system
Joints		
Endocrine	Kidneys	Lacrimal glands
Breast		
Uterus		

1- Lungs: - granulomas involve the interstitium rather than airspaces (around bronchioles and pulmonary venules and in the pleura)

- In 5% to 15% of cases, granulomas are replaced by fibrosis

2- Lymph nodes: a- hilar and paratracheal lymph nodes

b- peripheral lymph nodes: - occurs in one third of patients

- the node "nonmatted" and do not ulcerate (TB)

3- Skin:

- encountered in approximately 25% of patients

1- Erythema nodosum: - the hallmark of acute sarcoidosis

- consists of raised, red, tender nodules

- on the anterior aspects of the legs

2- painless subcutaneous nodules

3- raised painless plaques

4- lupus pernio:- indurated plaques

- in nose, cheeks, and lips

4- Eye and lacrimal glands:

- in the form of conjunctivitis, iritis, retinitis
- suppression of lacrimation (sicca syndrome)

- complicated by: corneal opacities, glaucoma, and total loss of vision

5- Salivary glands:
- parotitis (painful enlargement with xerostomia)

6- Spleen, liver: enlargement

7- CNS:
- Cranial nerves, and peripheral nerves can be involved
- 7th nerve facial palsy is most common
- Hereford's syndrome: facial palsy accompanied by fever, uveitis, and enlargement of the parotid gland

8- Heart:
- involved in about 5% of cases
- in the form of:
    1- conduction abnormalities (arrhythmias)
    2- cardiomyopathy
    3- sudden death

9- Musculoskeletal:
- acute polyarthritis with fever is common
- chronic destructive bone disease with deformity is rare
- polymyositis and chronic myopathy

10- Kidneys:

- granulomatous interstitial nephritis

Clinical Course: either:

- asymptomatic: discovered on routine chest X-ray (as bilateral hilar adenopathy)
- symptomatic: - peripheral lymphadenopathy, cutaneous lesions, eye involvement, splenomegaly, or hepatomegaly may be presenting manifestations
  - respiratory symptoms (shortness of breath, dry cough, vague substernal discomfort)
  - constitutional signs and symptoms (fever, fatigue, weight loss, anorexia, night sweats)

Diagnosis:

1- Imaging studies (CT scan, X-ray)

2- Biopsies: lung, lymph node,……

3- Hypercalcemia and hypercalciuria:

- not related to bone destruction
- caused by increased calcium absorption secondary to production of active vitamin D by the mononuclear phagocytes in the granulomas

- sarcoidosis is a diagnosis of exclusion (must rule out other granulomatous diseases)

Treatment:

- steroid therapy

Prognosis:

- 65% to 70% of affected individuals recover
- 20% develop permanent lung dysfunction or visual impairment
- 10% to 15% develop progressive pulmonary fibrosis and cor pulmonale

Thank you