Sarcoidosis

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- 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