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