

## **SARCOIDOSIS**

### **A Systemic Granulomatous Disorder**

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Sarcoidosis, is an inflammatory systemic autoimmune condition characterized by the formation of granulomas, which can cause fibrosis and interfere with organ function.

### **Sarcoidosis and Granulomas**

Sarcoidosis is an inflammatory systemic autoimmune disease first described in the late 1860s. Sarcoidosis is characterized by small lumps of cells known as granulomas. These granulomas, which are produced in response to inflammation, are granular in appearance, and can, normally, only be seen with the aid of a microscope.

Histologically, these granulomas are composed of epithelial cells and are not contained within capsules. When many granulomas arise in close proximity and clump together in one organ, they can interfere with the organ's function.

### **Who is Affected?**

Sarcoidosis affects people of all ages although it primarily targets young adults between the ages of 20 and 40 years. Sarcoidosis is most common in Northern Europeans and American blacks. The lifetime risk of developing sarcoidosis is highest among American blacks, affecting 2.4 percent of this population; Swedish women, affecting 1.6 percent; and Swedish men, affecting 1.15 percent.

### **Organs Affected by Sarcoidosis**

Sarcoidosis can occur in nearly any part of the body although some organs are affected more than others, and usually at least two organs are affected simultaneously. The most common organs affected by sarcoidosis include:

- \* Lungs, with imaging test abnormalities seen in 90 percent of patients with sarcoidosis
- \* Lymph Nodes, especially those found in the chest cavity
- \* Skin
- \* Eyes
- \* Liver

Other organs that may be affected by sarcoidosis, although less often, include:

- \* Spleen
- \* Brain
- \* Nerves

- \* Heart
- \* Tear Glands
- \* Salivary Glands
- \* Bones and Joints

Rarely, sarcoidosis can affect the following organs:

- \* Thyroid Gland
- \* Breasts
- \* Kidneys
- \* Reproductive Organs

### **Disease Course in Sarcoidosis**

Sarcoidosis has an active and an inactive phase. In the active phase, the formation of granulomas can cause symptoms, and it can produce scar tissue leading to fibrosis. In the inactive phase, inflammation subsides, and the granulomas remain stable in size or they may shrink. However, scar tissue may cause symptoms. Symptoms range from mild to severe, and in many cases symptoms resolve within a few years. Alternately, symptoms may persist, arising as flares periodically, and in some cases sarcoidosis gradually worsens over time, causing permanent organ damage.

Although the cause of sarcoidosis remains unknown, researchers suspect that a single provoking agent or a disordered immune system reaction may play a role in disease development. Genetic factors are also considered causative. Although granulomas may occur in response to hypersensitivity reactions, pneumonia and foreign body reactions, the pattern of granuloma formation in sarcoidosis differs in that it is progressive during the active phase.

### **Symptoms**

Symptoms vary, depending on the organ that is affected. Most commonly, the lungs are affected causing breathing disturbances and pulmonary fibrosis. Usually, symptoms emerge gradually although some symptoms may arise suddenly, including disturbed heart rhythms, arthritis in the ankles, and eye symptoms. Fever, weight loss, enlarged lymph nodes, and arthralgias are common early symptoms.

Skin lesions, such as plaques, nodules, and papules, are often seen in patients with chronic sarcoidosis and these lesions may occur in the eyes and noses. Erythema nodosum is more likely to be seen in European patients.

Granulomas of the liver are found on liver biopsies of most patients with sarcoidosis even when liver function tests remain normal. Granulomatous uveitis occurs in about 15 percent of patients with sarcoidosis and usually affects both eyes. Granulomatous uveitis that remains untreated may cause severe vision loss or a secondary form of glaucoma.

The heart is affected in 10-15 percent of patients. Myocardial granulomas may cause angina, heart failure or fatal conduction abnormalities.

Increased vitamin D production by sarcoid granulomas may cause increased calcium levels in the blood and urine (hypercalcemia and hypercalciuria respectively). The increased calcium production may cause the production of kidney stones and lead to conditions of both hypoparathyroidism and hyperparathyroidism. Diabetes insipidus may also occur.

Sarcoidosis can affect the male reproductive system, particularly the testes, causing male infertility and erectile dysfunction. Only rarely are the female reproductive organs affected.

### **Neurosarcoidosis**

The term neurosarcoidosis refers to the neurological manifestations of sarcoidosis. Symptoms vary widely and include,

- \* Facial palsy
- \* Muscle weakness and wasting
- \* Numbness in various parts of the body
- \* Excessive thirst
- \* Fatigue, Lethargy
- \* Headache
- \* Seizures
- \* Fever
- \* Memory Loss
- \* Irritability
- \* Agitation
- \* Depression
- \* Loss of taste
- \* Vocal changes,
- \* Hearing impairments
- \* Vision impairments

### **Treatment**

There is no known treatment for sarcoidosis although there have been several reports of success using antibiotic therapies. Corticosteroids or immunosuppressive drugs are also used to reduce symptoms in some cases.

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Sarcoidosis, National Heart Lung and Blood Institute, , accessed Sept 22, 2006.

Sarcoidosis Research Institute, Memphis, TN

Neurosarcoidosis, National Institute of Neurological Disorders and Stroke, accessed Sept 22, 2006.

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