



Medicine for Managers

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Sarcoidosis

Sarcoid, also called sarcoidosis, is a rare condition where small clumps of inflammatory cells develop and accumulate in a variety of areas of the body, most commonly in the lungs and lymph nodes. The annual incidence of the disease is about 7/100,000 and about 4,500 people were diagnosed in 2012. The incidence is higher in Ireland, Scotland and London and some people including Irish and Afro-Caribbean have a predisposition to it.

The collections of cells, also called **granulomas**, may develop in the lungs and lymph nodes, but also may affect the skin, heart, eyes and other organs.

The cause of the disease is unknown but it is believed to be an inflammatory immune response to an, as yet unknown, agent.

Such a response is essential for the body's normal defence and, when the invading agent is removed, the reaction dies down.

However, in sarcoidosis, it is thought that the body 'over-reacts' and actually attacks its own body proteins, resulting in the formation of the granulomas. As the granulomas build up in an organ, the structure and function can be affected and deteriorate if scarring develops.

The peak age of incidence is between 20 and 55. Women are more commonly affected than men. There is a link with family. It occurs more commonly in people where another family member already has the condition.

The disease has a variable course. For most people in whom the diagnosis is made, they do well without treatment over a period of months or years. The symptoms may appear suddenly and then disappear just as quickly.

For many, there are no symptoms when the disease is present and a diagnosis is made by chance when a chest X-ray is taken for another reason.

90% of those patients with sarcoidosis have lung changes, and the condition is termed **pulmonary sarcoidosis**. Those patients with symptoms most commonly have:

- Feeling of shortness of breath
- A cough which is often dry
- Wheezing
- Less commonly, chest pain

Other symptoms include

- Tiredness and fever
- Sweating
- Swollen, tender glands in the neck
- Skin rashes

- Painful red lumps on the shins, called erythema nodosum, may occur in between 10 and 20% of symptomatic sufferers
- Painful, red eyes
- Sometimes joint and muscle pain
- Occasionally a heart dysrhythm

The sarcoid may cause a variety of skin problems ranging from a purplish raised rash, areas of pigmented or pale skin, and skin nodules (especially associated with scars) to sores on the nose, ears or cheeks which may scar and become distorted.

The eye changes may be reddening, pain, itching and blurred vision.

Apart from the lung effects which can lead to scarring (**pulmonary fibrosis**) and the heart effects leading to abnormal rhythms, sarcoid can be associated with the formation of kidney stones and the development of granulomas in the brain can, on occasion, cause facial paralysis or fits.

Sarcoidosis may be difficult to diagnose, especially if the symptoms are mild or intermittent.

The disease may be mistaken for other disorders. In a patient with symptoms the usual types of tests are undertaken including blood tests, urinalysis, chest X-ray, ECG, CT scan and a number of other physical and blood tests.

Sarcoid still remains a disease where the diagnosis is considered a possibility after other causes have been excluded.



In the presence of granulomas, the diagnosis can be made by biopsy of a lesion. A sample of tissue is most commonly either taken from the lung through an endoscope or from the skin.

There is no cure for sarcoidosis and, as I have said, in many of the cases it resolves spontaneously.

Even in some of the patients diagnosed, there are either no symptoms or those that are present are mild and do not require any treatment. For patients with more severe or more widespread disease, the nature and distribution will govern the medical approach.

There are a range of drugs which may help in controlling and suppressing the disease:

- **Steroids.** They are a first-line treatment and can be taken orally, applied to skin lesions as a cream or into the eyes as drops.
- **Immunosuppressant Drugs** such as methotrexate or azathioprine can suppress the immune system
- **Tumour Necrosis Factor (TNF) drugs** may be effective in sarcoid resistant to other treatments.
- **Cardiac medication** may be necessary for patients with dysrhythmias or signs of heart failure or raised blood pressure

In extreme cases where there is extensive organ damage through granulomatous change in the lungs, liver or heart, organ transplantation may be considered.

As indicated, sarcoidosis affects different people in different ways

- It may last a few weeks or months and may require no treatment. Once the disease has remitted, it is very unlikely to recur.
- The chronic form may last for years but may not progress.

Most people with the disease do improve but the severe form with serious consequences occurs in only a small proportion of patients.

Sarcoidosis UK is a charity which exists to fund research into a cure for sarcoidosis.

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www.sarcoidosisuk.org

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