

# CASE REPORT OF PULMONARY SARCOIDOSIS

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## ABSTRACT:

Pulmonary sarcoidosis is a rare disease characterized by the growth of tiny collections of inflammatory cells (Granulomas) in any part of your body. Most commonly the lungs and lymph nodes. But it can also affect the eyes, skin, heart, and other organs. The cause of sarcoidosis is unknown, but experts think it results from the body's immune system responding to an unknown substance. Some research suggests that infectious agents, chemicals, dust, and a potential abnormal reaction to the body's own proteins (self-proteins) could be responsible for the formation of Granulomas in people who are genetically predisposed. A patient was admitted to the hospital with cough, breathlessness, headache, and pain in the lower limbs but the patient was diagnosed with pulmonary sarcoidosis. The patient was treated with drugs for a symptomatic cure and recovered from his condition and improved significantly.

**Keywords:** Pulmonary sarcoidosis, Granulomas, Immune system, Lungs, and Lymph nodes.

## INTRODUCTION:

Sarcoidosis was first described by Besnier et al. in 1889. It is a multi-system disease of unknown etiology characterized by the infiltration of various organs by non-necrotizing granulomas. Even if sarcoidosis remains a disease of unknown cause, the mechanisms underlying granuloma formation are better and better understood, including genetic susceptibility and environmental factors. <sup>(1)</sup> Around 50% of patients present with pulmonary complaints such as dyspnea on exertion, cough, and chest pain. <sup>(2)</sup> The disease occurs in both men and women with a slight predominance affecting women, and most patients are aged between 20 and 50 years. <sup>(3)</sup> Since the lungs are often involved, patients usually come to the clinic with lung complaints (such as shortness of breath, and cough). Some of the clinical manifestations of sarcoidosis have a poor prognosis, including treatment-resistant lung sarcoidosis. <sup>(4)</sup> The epidemiology of sarcoidosis and PH demonstrates that sarcoidosis-associated pulmonary hypertension (SAPH) is prevalent, highly morbid, and deadly in patients that are commonly seen by pulmonologists. Between 5.7 and 28.3% of all sarcoidosis patients develop SAPH, with a wide range of prevalence reported across several single-centrestudies. <sup>(5)</sup> Biomarkers and gene polymorphisms may provide help in assessing risk for sarcoidosis, predicting disease outcome, or assessing treatment response, but they still do not have a role in the routine diagnosis of sarcoidosis. <sup>(6)</sup> Sarcoidosis is characterised by a T-helper response in which CD4 lymphocytes and activated macrophages accumulate in affected organs, resulting in the formation of granulomas believed to develop in patients with genetic susceptibility after exposure to unidentified antigens and activating undifferentiated histiocytes into M2 phenotype macrophages; expansion and polarisation of CD4+ lymphocytes towards Th1, Th17, and Th17.1 cells; impairment of T regulatory cell (Treg) function; and the development of sarcoid granulomas with inflammation that release a broad spectrum of mediators, including cytokines, chemokines, and oxygen radicals were involved in aetiopathogenesis. <sup>(7)</sup> Hypocalcaemia is caused by increased conversion of vitamin D to the activated form (1,25 hydroxy vitamin D) by macrophages. Hypercalciuria may be present even in patients with normal serum calcium levels. Nephrolithiasis and nephrocalcinosis occur, as well as chronic kidney disease. <sup>(8)</sup> The diagnosis of sarcoidosis relies on the presence of noncaseating granuloma on histopathologic examination, compatible clinical presentation, and the exclusion of other causes of granulomatous inflammation. The only exceptions to the histopathologic requirement are stage I pulmonary sarcoidosis, which the presence of bilateral hilar adenopathy alone is generally considered sufficient for diagnosis after exclusion of other possible causes, and Löfgren syndrome (i.e., bilateral hilar adenopathy accompanied by erythema nodosum, fever, and arthritis). <sup>(9)</sup> In 57–88% of cases, fiberoptic bronchoscopy granulomas were found by means of mucosal or transbronchial biopsy. Lymphocytosis in bronchoalveolar lavage (BAL) is observed in 90% of cases, and a CD4+/CD8+ T lymphocyte ratio greater than 3.5 in half of the cases. Transbronchial needle aspiration makes possible valuable samplings of hilar and mediastinal lymph nodes. <sup>(9,10)</sup> Chest high-resolution computed tomography (HRCT) has better diagnosis accuracy than chest X-ray. The hallmark of pulmonary sarcoidosis is widespread micronodules with a typical peri lymphangitic distribution and a predominance in the middle and upper parts of the lungs. However, HRCT is not always necessary when a confident diagnosis can be made from typical clinical and radiographic features. By contrast, HRCT makes compelling diagnostic contributions in tricky cases and in detecting complications of lung disease. HRCT is also particularly useful in cases that are difficult to treat. <sup>(10)</sup>

## CASE REPORT:

A male patient was admitted to the male medicine ward in a government general hospital with the chief complaints of breathlessness and cough for one week. Previously the patient was diagnosed with pulmonary sarcoidosis (Stage II) by the help of CT scan findings.



**Figure 1:**

The physician doesn't know the exact reason for causing pulmonary sarcoidosis (maybe genetics). The patient got the treatment of syp ambroxol for cough and O<sub>2</sub> inhalation for breathlessness. Laboratory investigations were Total count-15,600/cum, Neutrophils-78%, Urea-21 mg/dl, serum creatinine-0.5 mg/dl, and total bilirubin-0.6 mg/dl remaining were normal.

**Outcomes and follow-up:**

After diagnosing the condition patient was advised to use O<sub>2</sub> inhalation for breathlessness and syp. Ambroxol for the cough to prevent the worsening of the condition. The patient also got antibiotics and corticosteroid treatment for pulmonary sarcoidosis. After 14 days of treatment, the patient showed significant improvement in his condition and got discharged.

**DISCUSSION:**

Sarcoidosis is a multi-system, granulomatous disease without a known etiology. However, it is characterized by a T-helper cell response to CD-4 lymphocytes and activated macrophages that accumulate in the affected organs.<sup>(11)</sup> Most studies suggest that the pathogenesis is related to an exaggerated immune response to an environmental factor, microbe, or antigen in a genetically susceptible individual.<sup>(12)</sup> Although the worldwide epidemiology of sarcoidosis is difficult to ascertain due to a large proportion of patients being asymptomatic, it has been estimated that 60 out of 100,000 adults in the United States will be affected by the disease.<sup>(13)</sup> More than 80% of these cases will be diagnosed between the ages of 20-50 with a second peak in incidence between 50-65 years of life<sup>(11,12)</sup> Females, non-smokers, and Blacks are more commonly diagnosed with the disease and 10% of cases will be familial.<sup>(11,13)</sup> Mortality has been estimated at between 2-5% secondary to pulmonary complications, while morbidity can be substantial due to poor outcomes in chronic sarcoidosis.<sup>(13)</sup> Up to one-third of patients are asymptomatic at their time of presentation with findings discovered incidentally.<sup>(15)</sup>

Clinical presentations most commonly include unexplained or persistent cough, dyspnea, or chest pain.<sup>(15,16)</sup> Treatment for sarcoidosis is recommended for those with active disease and who are symptomatic.<sup>(13)</sup> First-line treatment for those with symptomatic stages II or III is systemic corticosteroids and follow-up every three months. Corticosteroids are also recommended for those with serious extrapulmonary disease. Treatment with 20-40 milligrams of prednisone per day for four to six weeks and tapering slowly if the condition improves is the mainstay of therapy.<sup>(12)</sup> Response to treatment is monitored every three to six months using clinical response, pulmonary function testing, and CXR.<sup>(12, 16)</sup> The results of the study are in line with studies that have suggested a good prognosis among patients with sarcoidosis. Other low rates of development of respiratory impairment have been noted by other investigators as well.<sup>(17)</sup> The stage of pulmonary sarcoidosis at diagnosis was a strong predictor of chronic respiratory impairment in this cohort. This is consistent with studies on the radiographic progression of pulmonary sarcoidosis, as spontaneous resolution of radiographic abnormality is seen in 60%-80% of patients with stage I pulmonary sarcoidosis. Resolution was observed less frequently, approximately 30%-50% with stage II and III disease.<sup>(16, 17)</sup>

**CONCLUSION:**

Sarcoidosis is susceptible to encompassing numerous different clinical presentations. Whether it is symptomatic or not, or acute or not, sarcoidosis can involve variable organs with a diverse clinical impact, from benign to very severe and also enigmatic multisystemic disease, has many different faces. Every patient has his or her own story, and every sarcoidologist has a unique but complex approach to maintaining the disease. Although multisystemic involvement is characteristic of the disease and virtually any organ can be affected, the lungs are involved in more than 90% of patients who have sarcoidosis. The probability of sarcoidosis diagnosis varies from "definite" to only "possible" depending upon the presence of more or less characteristics, radioclinical and histopathological findings, and the epidemiological context. Radiographic features and lung function impairments are supportive in managing how serious the disease is in an individual patient but are not the parameters for a definite prognosis. Pleural involvement is a rare but serious presentation of this perplexing disease. The main differential diagnosis includes infections, especially tuberculosis, and malignancies, especially lymphoma. This highlighted the existence of distinct phenotypes of sarcoidosis with a non-random distribution of organ involvement, which are associated with gender, geographical origin, and socio-professional category. Currently, these phenotypes should be used to understand sarcoidosis not as a single disease but as various syndromes. Genetic and environmental determinants of such phenotypes have to be elucidated in future studies to better understand the pathophysiology of sarcoidosis and eventually to guide treatment.

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