

## **SARCOIDOSIS, A DIAGNOSTIC CHALLENGE CASE REPORT**

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***Bethânia Bertoldi Soares***

Instituto Hospital de Base do DF (IHBDF)  
Brasília, DF  
<https://orcid.org/0000-0001-9236-1329>

***Jessika Lopes Torquato***

Instituto Hospital de Base do DF (IHBDF)  
Brasília, DF  
<https://orcid.org/0000-0003-4287-1595>

***Ludmila Naves Marinho***

Instituto Hospital de Base do DF (IHBDF)  
Brasília, DF  
<https://orcid.org/0000-0002-2364-7523>

***João Daniel Bringel Rego***

Instituto Hospital de Base do DF (IHBDF)  
Brasília, DF  
<https://orcid.org/0000-0002-8604-9800>

***Jefferson Fontinele e Silva***

Instituto Hospital de Base do DF (IHBDF)  
Brasília, DF  
<https://orcid.org/0000-0001-5912-273X>

***Danilo Martins de Sá***

Instituto Hospital de Base do DF (IHBDF)  
Brasília, DF  
<https://orcid.org/0000-0001-8123-106X>

***Fabrcio Sanches Leornec Dias***

Instituto Hospital de Base do DF (IHBDF)  
Brasília, DF  
<https://lattes.cnpq.br/2006668395483815>

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***Pâmella Bertoldi Soares***

Santa Casa de Misericórdia de Ribeirão Preto  
Ribeirão Preto, SP  
<http://lattes.cnpq.br/6644287381336748>

***Thales Marques Correa***

Santa Casa de Misericórdia de Ribeirão Preto  
Ribeirão Preto, SP  
<http://orcid.org/0000-0002-3147-6437>

***Tatiana Amabile Orben***

<http://lattes.cnpq.br/0849159721110909>

**Abstract: Introduction:** Sarcoidosis is defined as a multisystem granulomatous disease of unknown cause. It affects the lung in 90% of cases, but it can affect any part of the body. It can be self-limiting or chronic, with progression to terminal fibrosis. Its diagnosis is based on the compatible clinical-radiological correlation, in addition to the histopathological finding of non-caseating granuloma. **Objective:** To report a case of sarcoidosis from initial suspicion to diagnostic confirmation and treatment. **Description of case:** This is a male patient with a dry, dragging cough and unintentional weight loss. After investigation, diagnosis was made. **Conclusion:** Because it is a disease with diverse symptoms, including asymptomatic individuals or with unspecific symptoms, diagnosis is a challenge in clinical practice. The prognosis is usually favorable, but up to 25% of patients develop severe disability. Therefore, it becomes evident the importance of an early diagnosis for adequate management and continuous and multidisciplinary follow-up to avoid a chronic course. The prognosis is usually favorable, but up to 25% of patients develop severe disability. Therefore, it becomes evident the importance of an early diagnosis for adequate management and continuous and multidisciplinary follow-up to avoid a chronic and progressive course of the disease.

**Keywords:** Sarcoidosis, Lymph node enlargement, Granulomatosis.

## **INTRODUCTION**

Sarcoidosis is defined as a multisystem disease of unknown etiology. It is characterized by the presence of non-caseating granulomatous inflammation. It has variable presentation, progression and prognosis, with pulmonary manifestations present in up to 90% of patients, of which 20% to 25% develop permanent functional impairment. In addition to the lung, it can affect any part of

the body (MELLO; MARCHIORE; CAPONE, 2011).

Also known as Boesnier-Boeck-Schaumann disease, it is known that most patients, at the time of diagnosis, are aged between 20 and 60 years. It is also important to highlight its prevalence over the population residing in tropical regions and under temperate climates (GOMAR *et al.*, 2021).

It is usually a self-limiting disease for most of its carriers, which cure or stabilize without the use of immunosuppressants or anti-inflammatory agents. In some cases, extensive and irreversible pulmonary fibrosis may develop, which leads to progressive deterioration of respiratory function (LOPES; COSTA; RUFINO, 2013).

According to Deubelbeiss *et al.* (2010), its clinical presentation is quite varied, ranging from asymptomatic, with only findings on chest radiography, to the form with involvement of multiple organs such as skin (25-35% of cases), eyes, heart, liver, kidneys, salivary glands and lymphoid system.

Regarding epidemiology, the highest prevalence occurs in countries in northern Europe, with a record of 5-40 cases/100,000 inhabitants. In Brazil, the incidence is estimated at 10/100,000 inhabitants. Studies cited by Lopes, Costa and Rufino (2013) clearly demonstrate the existence of variability of sarcoidosis in relation to gender, age and ethnicity, pointing out that, in addition to the possible preference for some ethnic groups, the pathology also presents itself in a more aggressive way in certain populations. The authors also point out that in a case-control study with 736 cases of sarcoidosis evidenced by biopsy, it was shown that, although the disease is more prevalent in white Americans (53% of the patients in the study) than in blacks, the involvement of extrathoracic sarcoidosis was more common in the latter group.

Commonly presented respiratory manifestations include cough, dyspnea, and chest pain, which may also be associated with asthenia, malaise, fever, or weight loss. On physical examination, fevers are uncommon and wheezing may be present when there is endobronchial involvement or traction bronchiectasis (BRANCO *et al.*, 2016).

Regarding the diagnosis of sarcoidosis, it is based on the presence of a compatible clinical-radiological correlation, in addition to the histopathological finding of non-caseating granulomas and exclusion of other granulomatous diseases. The fact is that all people with a suspected diagnosis of sarcoidosis must undergo a clinical evaluation, which includes a complete clinical history and physical examination (BRANCO *et al.*, 2016).

Treatment is aimed at preventing and controlling organic deterioration, symptomatic relief and improving quality of life. Corticosteroids are considered to be the first-line treatment for the disease. Alternative therapies with antimalarials, immunosuppressants and cytotoxic agents have also been used in the treatment of sarcoidosis (MYERS; TAZELAAR, 2008).

Based on the above, the present report aims to describe the case of a 53-year-old male patient diagnosed with sarcoidosis after presenting with dyspnea on moderate exertion, dry cough and unintentional weight loss.

### **CASE REPORT**

This is a male patient, 53 years old, car painter, hospitalized for investigation of wasting syndrome and interstitial disease.

On admission, he reported a dry cough and unintentional weight loss (about 20 kg) over a year, associated with progressive dyspnea, sporadic dysuria and difficulty initiating urination. He denied fever, sweating, hemoptysis, wheezing, among other

symptoms.

He had diabetes mellitus and systemic arterial hypertension as comorbidities. Ex-smoker patient (smoking history of 10 packs/year, abstainer for 25 years) and ex-alcoholic (abstaining for 4 years). He denied known lung diseases and a family history of neoplasia. He had previously been hospitalized with the same symptoms.

With an occupational history, he reports working with automotive painting for 30 years, without using personal protective equipment.

On physical examination, he was emaciated, had creptations in the upper third of the HTE and in bases, SpO<sub>2</sub>: 93% in room air, eupneic, without respiratory distress. The abdomen was hollow and painless on palpation. He presented xeroderma all over the body, especially in the lower limbs.

He had HIV-negative genexpert and serology, echocardiogram showing minimal anterior pericardial effusion, LV diastolic dysfunction and asynchronous movement of the interventricular septum, in addition to a chest tomography with diffuse pulmonary alterations, ground-glass infiltrate, reticular opacities suggestive of NSIP, small nodules of random distribution and mediastinal lymph nodes increased in number and size.

Figure 1

Figure 2

In view of these findings, pleuroscopy with biopsy of the lung parenchyma was performed, whose preliminary result suggested the presence of granulomatous disease. Prednisone 40 mg/day was started and discharged for outpatient follow-up.

The patient returned for consultation 30 days after discharge, with improvement in cough and dyspnea, with the definitive anopathological result showing a non-necrotizing granulomatous chronic inflammatory process in the lung. In view of the clinical improvement, it was decided to

start weaning from corticosteroids.

Nine months after the start of treatment, a new chest tomography was performed, which showed improvement in the ground glass and perilymphatic micronodules and maintenance of fibrosis with traction bronchiectasis predominating in the apices.

Figure 3

The patient maintained regular follow-up at the pulmonology outpatient clinic and, after one year of treatment, the corticosteroid was discontinued, with complete resolution of the previous cough, dyspnea on exertion, weight gain with return to normal and good diet acceptance.

At that time, the patient had already managed to return to his usual activities.

## DISCUSSION

We study the case of a patient diagnosed with sarcoidosis after presenting with dyspnea, dry cough and weight loss. Sarcoidosis is a multisystem inflammatory disease of unknown etiology, characterized by the formation of non-caseating granulomas in Organs involved organs, predominantly lungs and intrathoracic ganglia. The presence of cutaneous involvement occurs in about 25-35% of the cases, being an early finding, constituting an accessible and safe place for histological study. Skin lesions can have various presentations, including papules, nodules, plaques and infiltrated scars (BRANCO *et al.*, 2016).

It is a pathology that presents a well-structured core of knowledge from different points of view, whether of an epidemiological, clinical, laboratory, imaging, immunological or therapeutic measures nature, however, it often exhibits a periphery, sometimes wide, of variety. of presentation and also of ignorance (BETHLEM, 2005).

Regarding the etiopathogenesis of sarcoidosis, although its etiology is

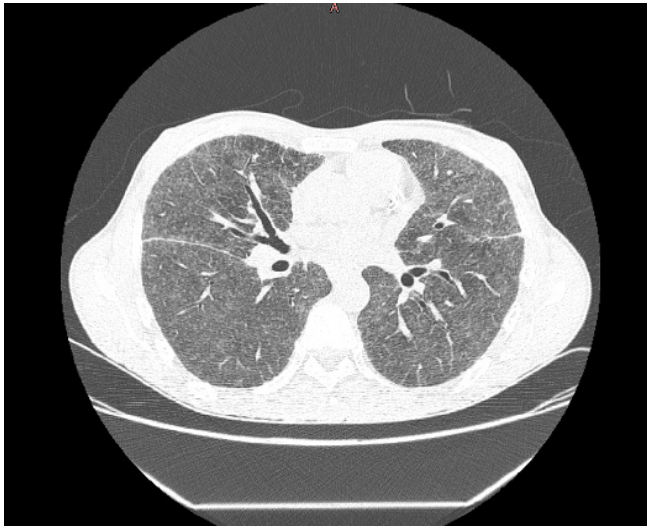


Figure 1: VF opacities diffusely distributed throughout the lung parenchyma. mild paraseptal emphysema



Figure 2: Thickening of the interlobular septa

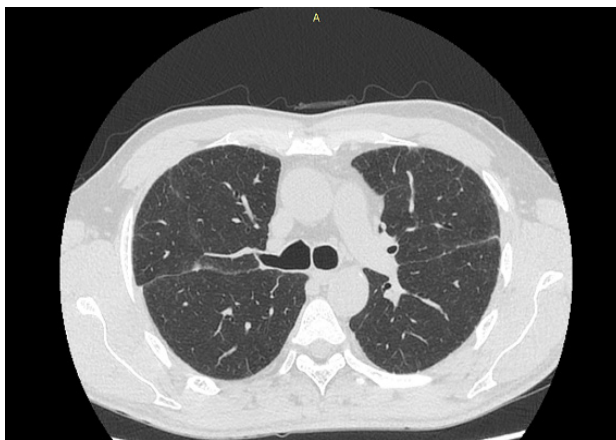


Figure 3: Perilymphatic micronodules in the upper lobes, associated with discrete peripheral ground glass

unknown, several advances have been made in its understanding. It has currently been hypothesized that environmental exposure (organic or inorganic agents) in genetically predisposed hosts greatly contributes to the activation of the immune response, granulomatous inflammation and secondary fibrotic tissue injury. (BRANCO *et al.*, 2016).

According to Gomar *et al.*, (2021) with bilateral and asymmetric manifestation, granulomas arise from the union of inflammatory cells combined with changes in the physiological immune conformation. From the progression of the disease, the development of fibrosis around the lesion sounds relevant.

In approximately half of the cases, the diagnosis is established “accidentally” by imaging changes in asymptomatic individuals.

However, although radiological findings such as hilar adenopathies are characteristic, additional tests are mandatory to differentiate it from other granulomatous diseases such as tuberculosis.

The hypothesis that environmental agents are involved in the etiopathogenesis of sarcoidosis is supported by some researchers who suggest a high risk in individuals exposed to insecticides, in moldy environments and in agricultural jobs. Furthermore, this hypothesis is based on its occurrence in certain professional groups, including firefighters, ship crews, wood cutters, mechanics, health professionals and photocopiers (BAUGHMAN; LOWER; BU BOIS, 2003; COX, ALLEN, JUDSON, 2005; NUNES; SOLER, VALEVRE, 2005).

The findings of the authors above are in line with our case report, since the patient under study reported working with automotive paint for 30 years, without using personal protective equipment, and being exposed to toxic material for a long period of time.

For Branco *et al.* (2016), among the possible

antigens of the pathology under study, there are infectious, organic and inorganic agents. Sarcoidosis granuloma is characterized by a chronic inflammatory reaction formed by the accumulation of epithelioid cells, monocytes, lymphocytes, macrophages and fibroblasts, and at the pulmonary level, granulomas are most commonly found in the alveolar septa, bronchial walls and pulmonary arteries or veins.

The most frequent respiratory manifestations include cough, dyspnea and chest pain, which may be associated with asthenia, malaise, fever or weight loss. These findings are also in line with the patient under study, who reported weight loss, dry cough and dyspnea. On physical examination, fevers are uncommon and wheezing may be present when there is endobronchial involvement or traction bronchiectasis (BRANCO *et al.*, 2016).

For Vizza *et al.*, (2017), for the diagnosis, other granulomatous diseases must be excluded. Bilateral hilar adenopathy is the most common abnormality, and if sarcoidosis is suspected, chest radiography must be the first test if it has not already been done. Radiographic appearance tends to approximately predict the probability of spontaneous remission. The fact is, the exclusion of other diagnoses is crucial, especially if radiographic signs and symptoms are minimal, given that many other diseases and processes can cause granulomatous inflammation. These are the statements by Gomar *et al.* (2021):

Clinically, Besnier-Boeck-Schaumann disease is usually diagnosed in an asymptomatic context. However, between 15 and 40% of patients affected by the present disease suffer from symptoms such as cough and dyspnea; On physical examination, auscultation of crackling rales is recurrent in less than 20% of those affected. From the point of view of diagnosing sarcoidosis, a

pathological study with biopsy is usually necessary in order to determine the presence of the disease – which justifies the late diagnosis of this disease; in some cases, on the other hand, the importance of imaging tests for pathological confirmation is relevant (GOMAR et al., 2021, p. 198).

Vizza et al. (2017) state that pulmonary symptoms range from none to cough, dyspnea on exertion, and, rarely, respiratory or other organ failure. For the authors, initially, the diagnosis is assumed due to pulmonary involvement, confirmed by chest radiography, biopsy and exclusion of other causes of granulomatous inflammation. The prognosis is excellent for limited disease but poor for more advanced disease.

Regarding treatment, corticosteroids are considered the first-line treatment for sarcoidosis. There are still alternative therapies with anti-malarials, immunosuppressants and cytotoxic agents that have also been used in the treatment of sarcoidosis. In the case report, the patient used prednisone 40mg/day for two weeks, followed by 30mg/day, and 20mg/day until he was weaned. Corticosteroids reduce systemic inflammation in most patients and, therefore, can delay or prevent organ damage (IANNUZZI et al., 2007).

For Spagnolo (2018), generally, the disease treatment protocol requires the therapeutic use of specific medication only to those that describe symptomatic manifestation with damage to the respiratory system such as corticosteroids (SPAGNOLO, 2018).

Regarding the prognosis, although spontaneous remission is common, the severity and manifestations of the disease vary, and many patients, at some point, require therapy with corticosteroids during the course of the disease. Therefore, serial monitoring to verify evidence of relapse is indispensable. Almost two-thirds of patients with the condition eventually achieve remission with little or no sequelae, and about 50% of

patients who develop spontaneous remission do so within the first 3 years after diagnosis; less than 10% of these patients relapse after 2 years. Those who do not develop remission within 2 to 3 years are likely to have chronic disease (VIZZA et al., 2017).

## CONCLUSION

From the analysis and development of the case report involving sarcoidosis, it was possible to know more deeply its epidemiological, symptomatological and diagnostic aspects.

The symptomatology of sarcoidosis is very wide, ranging from asymptomatic individuals or individuals with nonspecific symptoms, which makes the diagnosis even more challenging. The prognosis is generally favorable, especially in patients whose diagnosis and management are established early. Only 10% develop severe and irreversible disability.

Therefore, it becomes evident the importance of a continuous and multidisciplinary follow-up to avoid a more chronic and progressive course of the disease, in addition to the emergence of possible complications.

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