Case Report

An Unusual Presentation of Sarcoidosis: The Great Mimicker

Sunil Kumar, Ajay Garg, Sheena Taneja, Ayushi Chander, Harsh Vyas

Department of Respiratory Medicine, Pacific Medical College and Hospital, Udaipur, Rajasthan, India

Abstract

Sarcoidosis is a very well-known disease for its variable clinical and radiological presentation. Thoracic involvement is the commonest manifestation of sarcoidosis followed by skin. We present here an interesting case report of an old-age patient with an unusual presentation of sarcoidosis. He was previously treated for leprosy based on skin biopsy with partial relief only. The unusual things in this patient were clinical and radiological findings. Clinically, he had only complaints of weight loss which had not been reported earlier; second, the skin lesions were not typical. The unusual radiological things were a disproportionate distribution of radiological lesions and predominant consolidation. Lower lobe consolidation in sarcoidosis is rarely reported, which was present in this patient. The perilymphatic micronodules were more concentrated on the right upper lobe as compared to the left upper lobes. Based on this case report we cannot deny the wrong diagnosis of leprosy because both diseases share common clinical and histopathological features.

Keywords: *Consolidation in sarcoidosis, sarcoidosis, unusual sarcoidosis*

Received: 20-06-2023
Revised: 07-07-2023
Accepted: 23-07-2023
Published: 04-12-2023

Introduction

Sarcoidosis is an idiopathic multisystem granulomatous disease with varying clinical presentations and an unpredictable clinical course.[1] In sarcoidosis, around 95% of patients have thoracic involvement, 50% have extrathoracic manifestations and 2% have unaccompanied extrathoracic sarcoidosis.[2] Skin involvement is the most common extrathoracic manifestation of sarcoidosis, which can be present in 20%–40% of individuals.[3] Around half of the patients with pulmonary sarcoidosis are symptomatic and usually present with dry cough, dyspnea, and nonspecific systemic constitutional symptoms such as fever, malaise, anorexia, and weight loss.[2] Thoracic sarcoidosis remains asymptomatic and is diagnosed incidentally on chest imaging in up to 50% of patients.[1-6]

A diagnosis of sarcoidosis is established from compatible clinical and radiologic findings, confirmed by a biopsy showing noncaseating epithelioid granulomas in more than one organ and the exclusion of granulomatous disorders of known cause. The diagnostic yield of transbronchial lung biopsy is estimated to be >40% even in patients with a Stage I chest radiograph and approaches 80% in the presence of lung infiltrates.[7] Transthoracic tru-cut biopsy has largely replaced the open lung biopsy in peripheral lesions, especially in large masses, which can be present in up to one-quarter of pulmonary sarcoidosis.[8] Here, we report an unusual and interesting patient with pulmonary sarcoidosis, which presented to us with only complaints of weight loss and the right lower lobe consolidation.

Case Report

A 68-year-old, never-smoker, non-alcoholic male was referred to us from the oncology department. He visited the oncologist with complaints of weight loss for the last 3 months and a fear of malignancy since his wife had been diagnosed with lung malignancy a few months earlier by the same oncologist. The oncologist suggested a few blood investigations and a *fluorodeoxyglucose* positron emission tomography-computed tomography (PET-CT). The PET-CT scan revealed

Address for correspondence: Dr. Sunil Kumar, Department of Respiratory Medicine, Pacific Medical College and Hospital, Udaipur - 313 001, Rajasthan, India.
E-mail: dr.sunil2004@gmail.com

How to cite this article: Kumar S, Garg A, Taneja S, Chander A, Vyas H. An unusual presentation of sarcoidosis: The great mimicker. J Adv Lung Health 2024;4:35-9.

Access this article online

Quick Response Code:

Website: www.jalh.org

DOI: 10.4103/jalh.jalh_23_23

a hypermetabolic area of collapse consolidation in the basal segment of the right lower lobe measuring 39.8 mm × 72.3 mm × 37.6 mm [Figure 1]. The lung images also showed disproportionately distributed perilymphatic nodules, predominantly on the right side and upper lobes. A few subcentimeter size lymph nodes were also seen in the right upper and the left lower paratracheal, prevascular, and para-aortic regions. The largest nodes were paraaortic with a size of 12.5 mm × 9.2 mm [Figure 2]. The PET-CT did not find any other definite hypermetabolic focal lesion in the body. A diagnosis of the right lower lobe pneumonia was made and the patient was started on empirical antibiotics by the oncologist. The patient was referred to us because of no improvement with antibiotics.

In his past medical history, he was diagnosed with tuberculoid leprosy and took treatment for 7 years with partial improvement in skin lesions. The basis of diagnosis was a histopathological examination of tissue biopsy from the left cheek skin nodule that revealed two noncaseating granulomas in the deep dermis in which one was noted surrounding the nerve bundle. He had no history of any chronic respiratory diseases and anti-tubercular treatment. He was nondiabetic and normotensive. His SARS CoV-2 reverse transcription polymerase chain reaction and HIV tests were negative.

Physical examination reveals diffuse erythema over the face with telangiectasia and a few papules present over the maxillary area, which worsens on exposure to the sun [Figure 3]. The physical signs such as pallor, icterus, cyanosis, clubbing, and lymphadenopathy were absent. On respiratory examination, a few crackles and bronchial sounds were heard over the right infrascapular region. His complete blood count, liver function tests, renal function tests, serum angiotensin-converting enzyme (ACE) levels, routine urine examination, and electrocardiogram were within the normal limits. The serum calcium was raised. His induced sputum was also negative for gram stain, aerobic culture, acid-fast bacilli, and GeneXpert.

We revised the antibiotic and suggested tissue biopsy from the peripheral lesion. The tissue biopsy showed multiple small noncaseating granulomatous lesions with giant cells, surrounded by fibrosis and lymphocytes. Asteroid bodies were seen, but Schaumann bodies were not seen. A diagnosis of sarcoidosis was established and the patient started on steroids.

The patient was kept under close observation with regular follow-ups. On subsequent follow-ups, the patients showed excellent clinical improvement in the form of weight gain and resolution of skin lesions. The serum calcium level became normal by the 6th week. The skin lesions get improved [Figure 4]. A repeat computed tomography was done after 2 months that showed complete resolution of perilymphatic nodules and opacity with few residual fibrotic bands [Figures 5 and 6].

Discussion

Sarcoidosis is known for its wide range of clinicopathological presentations and is often termed the "great mimicker."[9] The patients of sarcoidosis may be asymptomatic or may have a range of minimal symptoms to life-threatening organ involvement. The presentation may be acute, subacute, or indolent. Although systemic constitutional symptoms such as fever, fatigue, malaise, and weight loss may be seen in more than 50% of patients and even patients may present with only systemic symptoms, isolated weight loss has never been reported as a presentation of pulmonary sarcoidosis.[10]

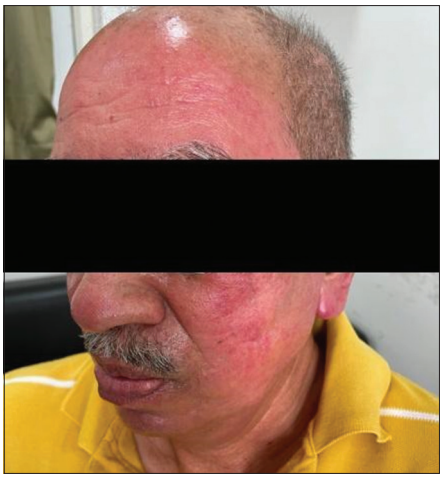

Figure 1: Skin lesions before the treatment. Note the malar area and forehead

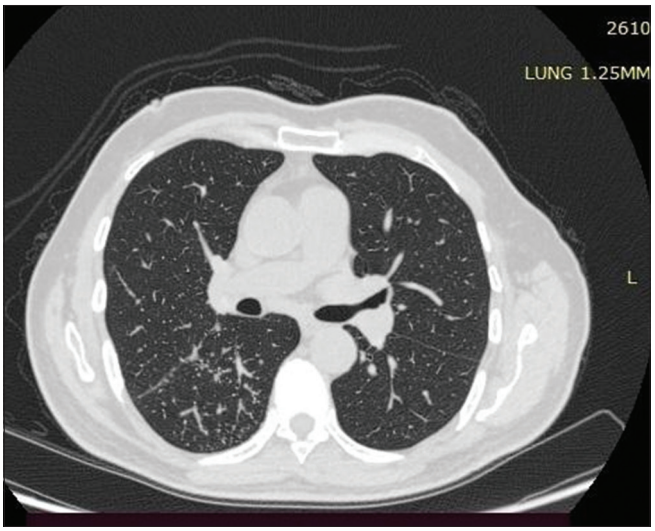

Figure 2: Chest computed tomography before the treatment. Note that the nodules are more prominent on the right side

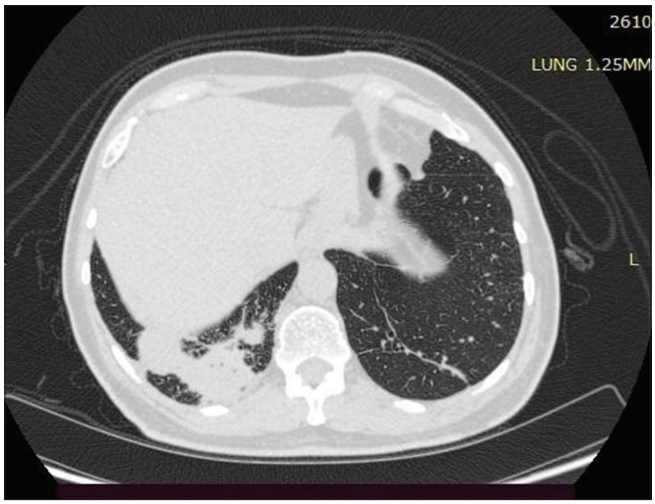

Figure 3: Chest computed tomography before the treatment. Note the right lower lobe consolidation

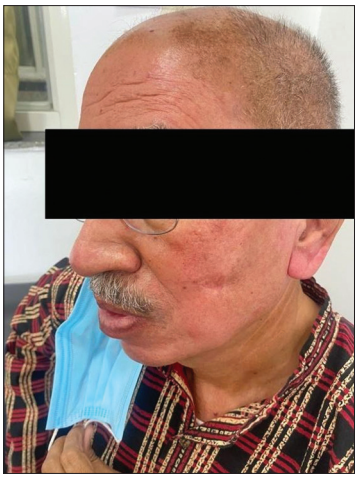

Figure 4: At 2 months of treatment. Note the improvement in malar and forehead skin lesions

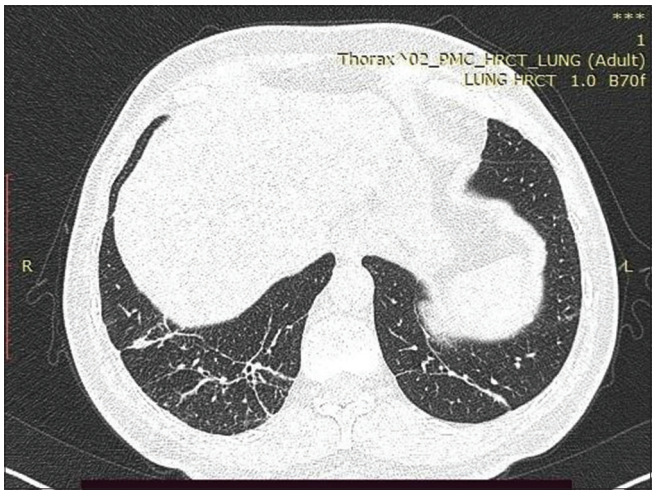

Figure 5: At 2 months of treatment. Note the resolution of nodules. HRCT: High-resolution computed tomography

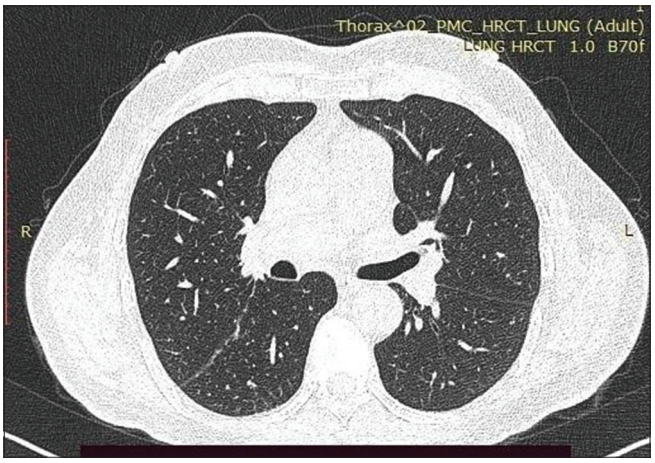

Figure 6: At 2 months of treatment. Note the complete resolution of consolidation and a few residual fibrotic bands. HRCT: High-resolution computed tomography

This unusual clinical presentation distracted the initial workup in our patient.

The typical skin lesions in sarcoidosis are described as plaques or subcutaneous nodules. The plaques are typically located around the hairline, eyelids, ears, nose, and extensor surfaces of the arms and legs. Other skin lesions include scar granuloma, tattoo granuloma, alopecia, and lupus pernio which is a disfiguring form of cutaneous sarcoidosis of the face. In acute sarcoidosis, erythema nodosum, which is pathologically nongranulomatous panniculitis, may be seen.[11] In our patients, the skin lesions were limited to only the maxillary area and forehead. The distinct thing about skin lesions in our patient was photosensitivity which is rarely reported in sarcoidosis.[11,12]

The chest radiograph is abnormal in more than 90% of patients in some stages of the disease.[13] Involvement of thoracic lymph nodes is present in more than 75% of cases, while lung parenchymal involvement occurs in more than 60% of cases.[14] Lymphadenopathy is usually bilateral and typically described as potato nodes. The lung parenchyma is usually involved bilaterally and symmetrically in the form of perilymphatic micronodules in the upper and middle lobes.[10] Atypical radiological findings may be present in as high as 30% of cases of sarcoidosis.[9] Unilateral and asymmetrical involvement of lymph node and lung parenchyma, necrosis and citation of nodules, large opacity, ground glass opacity, miliary nodule, and pleural effusion are considered unusual radiological findings in sarcoidosis. Although a frank consolidation is reported in up to 12% of patients with sarcoidosis, predominant lower lobe involvement is rarely reported.[15,16] Patients with lower lobe sarcoidosis often have dyspnea, eye lesions, and skin involvement and usually had a normal serum ACE level.[15] In our patient, we found lower lobe consolidation as the predominant radiological finding,

which is quite a rare occurrence. Although our patient had skin involvement and normal serum ACE level, the absence of dyspnea and eye lesions further makes this patient atypical. A normal serum ACE level may be found in sarcoidosis if the diagnosis is made in an earlier stage and the inactive state of the disease. In our patient, the normal serum ACE level was probably because of the long duration of the diseases. There is one study in which the author found an inverse relationship between the chronicity of sarcoidosis and serum ACE level.[17]

Histopathologically cutaneous sarcoid and tuberculoid leprosy are characterized by noncaseating granuloma. Both these diseases share common clinical and histopathological findings that can mimic each other and rarely coexist together.[18,19] Granulomas predominantly in adnexal and neural distribution, replacing the nerves and localized within sweat gland glomeruli, are predictive of tuberculoid leprosy. Fibrinoid necrosis, dermal fibrosis, back-to-back distribution of the granulomas, presence of atypical giant cells and plasma cells, greater number of conventional giant cells, and spared nerves beside the granuloma go in favor of cutaneous sarcoid.[20]

Previously, this patient was diagnosed with tuberculoid leprosy. The histopathology was examined by two pathologists. Although both pathologists identified granuloma surrounding the nerve bundle, we cannot deny the possibility of a wrong diagnosis. Further, we emphasize the role of lung biopsy in a diagnostic dilemma. At the first instance on clinicoradiological features, this case seems to be of infective lung pathology, but the lack of other laboratory findings was discordant. Sarcoidosis was not on the list of differentials because of atypical clinicoradiological presentation. The transthoracic lung biopsy, thankfully, ruled out malignancy and established the diagnosis of sarcoidosis. Lower zone lung tuberculosis could be one close differential for this patient, but the presence of asteroid bodies makes the diagnosis of sarcoidosis.

Although sarcoidosis is considered a disease of unknown cause, a few researchers have suggested the possible etiological role of *Mycobacterium tuberculosis* and atypical Mycobacteria in sarcoid.[21] *Mycobacterium leprae* had never been suggested as an etiological agent for sarcoidosis. Since leprosy and sarcoidosis share common immunopathogenesis, the possible role of *M. leprae* cannot be denied based on this case report.

Conclusion

The patients with sarcoidosis do not always present with typical clinical and radiological features. Weight loss alone may be the only clinical presentation of sarcoidosis. Disproportionate radiological lesions and lower lobe consolidation is possible in sarcoidosis. The diagnosis of sarcoidosis is aided by an understanding of the pattern of organ involvement and may ultimately require tissue confirmation through a biopsy of granuloma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Acknowledgments

The author would like to acknowledge the patient.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Lynch JP 3rd, Kazerooni EA, Gay SE. Pulmonary sarcoidosis. Clin Chest Med 1997;18:755-85.
2. Jain R, Yadav D, Puranik N, Guleria R, Jin JO. Sarcoidosis: Causes, diagnosis, clinical features, and treatments. J Clin Med 2020;9:1081.
3. Mangas C, Fernández-Figueras MT, Fité E, Fernández-Chico N, Sàbat M, Ferrándiz C. Clinical spectrum and histological analysis of 32 cases of specific cutaneous sarcoidosis. J Cutan Pathol 2006;33:772-7.
4. Baughman RP, Teirstein AS, Judson MA, Rossman MD, Yeager H Jr., Bresnitz EA, *et al.* Clinical characteristics of patients in a case control study of sarcoidosis. Am J Respir Crit Care Med 2001;164:1885-9.
5. Lynch JP 3rd, Ma YL, Koss MN, White ES. Pulmonary sarcoidosis. Semin Respir Crit Care Med 2007;28:53-74.
6. Judson MA, Boan AD, Lackland DT. The clinical course of sarcoidosis: Presentation, diagnosis, and treatment in a large white and black cohort in the United States. Sarcoidosis Vasc Diffuse Lung Dis 2012;29:119-27.
7. Bilaçeroğlu S, Perim K, Günel O, Cağirici U, Büyükşirin M. Combining transbronchial aspiration with endobronchial and transbronchial biopsy in sarcoidosis. Monaldi Arch Chest Dis 1999;54:217-23.
8. Brauner MW, Lenoir S, Grenier P, Cluzel P, Battesti JP, Valeyre D. Pulmonary sarcoidosis: CT assessment of lesion reversibility. Radiology 1992;182:349-54.
9. Criado E, Sánchez M, Ramírez J, Arguis P, de Caralt TM, Perea RJ, *et al.* Pulmonary sarcoidosis: Typical and atypical manifestations at high-resolution CT with pathologic correlation. Radiographics 2010;30:1567-86.
10. Grippi MA, Antin-Ozerkis DE, DelaCruz CS, Kotloff RM, Kotton CN, Pack AI. Systemic sarcoidosis. In: Chen ES, editor. Fishman's Pulmonary Diseases and Disorders. 6th ed. New York: Mcgraw Hill; 2023. p. 891-914.
11. Gangopadhyay A, Das JK, Sengupta S. Sarcoidosis with photosensitive lesions: A rare variant. Indian J Dermatol

2009;54:90-2.
12. Mahajan VK, Sharma NL, Sharma RC, Sharma VC. Cutaneous sarcoidosis: Clinical profile of 23 Indian patients. Indian J Dermatol Venereol Leprol 2007;73:16-21.
13. Ungprasert P, Ryu JH, Matteson EL. Clinical manifestations, diagnosis, and treatment of sarcoidosis. Mayo Clin Proc Innov Qual Outcomes 2019;3:358-75.
14. Miller BH, Rosado-de-Christenson ML, McAdams HP, Fishback NF. Thoracic sarcoidosis: Radiologic-pathologic correlation. Radiographics 1995;15:421-37.
15. Avital M, Hadas-Halpern I, Deeb M, Izbicki G. Radiological findings in sarcoidosis. Isr Med Assoc J 2008;10:572-4.
16. Matsui Y, Akagawa S, Masuda K, Yamato A, Ohshima N, Matsui H, *et al.* Nine cases of pulmonary sarcoidosis predominantly affecting the lower lung fields. Nihon Kokyuki Gakkai Zasshi 2010;48:883-91.
17. Kahkouee S, Samadi K, Alai A, Abedini A, Rezaiian L. Serum ACE level in sarcoidosis patients with typical and atypical HRCT manifestation. Pol J Radiol 2016;81:458-61.
18. Padiyar S, Kharkele R, Telugu RB, Rima S, Mathew J. A tale of two great mimickers: Leprosy or sarcoidosis? A case-based review. Indian J Rheumatol 2021;16:345-8.
19. Patil D, Sharda T, Krishnappa R, Sumathy TK. Co-existence of lepromatous leprosy and subcutaneous sarcoidosis: A rare paradoxical association? Lepr Rev 2022;93:413-19.
20. Utino FL, Damiani GV, Garcia M, Soares TC, Stelini RF, Velho PE, *et al.* Histomorphometric approach to differentiate skin lesions of tuberculoid leprosy from sarcoidosis. J Cutan Pathol 2018;45:111-7.
21. Brownell I, Ramírez-Valle F, Sanchez M, Prystowsky S. Evidence for mycobacteria in sarcoidosis. Am J Respir Cell Mol Biol 2011;45:899-905.