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CASE REPORT

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Cystic bronchiectasis in sarcoidosis

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Key Clinical Message

Pulmonary sarcoidosis can manifest in different radiologic patterns. Typical manifestations in high-resolution computed tomography are bilateral perihilar lymphadenopathy, micronodules, and fibrotic changes. Atypical manifestations are mass-like or alveolar opacities, honeycomb-like cysts, miliary opacities, tracheobronchial involvement, and pleural disease. Cystic bronchiectasis in pulmonary sarcoidosis is rare, with only a few reported cases in the literature. We present another case of cystic bronchiectasis with a honeycomb-like pattern in pulmonary sarcoidosis and with cardiac involvement. This case was presented as an abstract poster at the American Thoracic Society conference in 2022.

KEYWORDS

acute medicine, general medicine, radiology & imaging, respiratory medicine

1 | CASE REPORT

A 64-year-old man with no past medical history came to the emergency department complaining of progressively worsening shortness of breath for 6 months, associated with a productive cough for 2 days. History was remarkable for a weight loss of 25 lbs in 3 months. Physical examination revealed a cachectic male in respiratory distress with low oxygen saturation (86% on room air), which improved with supplemental oxygen. On auscultation, inspiratory crackles and expiratory wheezes were heard. An electrocardiogram showed a first-degree atrioventricular block. Chest radiograph (Figure 1) showed bilateral hilar adenopathy with a reticular consolidation pattern suggestive of interstitial lung disease. A high-resolution CT of the chest (Figures 2–4) revealed severe traction bronchiectasis of the upper lobes with multiple cysts in a honeycomb pattern, central and peripheral nodules, and mediastinal lymphadenopathy. Serological studies revealed eosinophilia of 7,22,000 cells/microliter, elevated angiotensin-converting enzyme (103 units/L), and IgG (2320 mg/dL) levels. Bronchoscopy showed a large conglomeration of lymph nodes at station 7, and pathology from a transbronchial biopsy revealed non-caseating epithelioid granulomas in the alveoli, confirming the diagnosis of pulmonary sarcoidosis. The patient was started on steroid therapy with improvement in symptoms.

2 | DISCUSSION

Sarcoidosis is a multisystem disorder characterized by the formation of granulomas of immune significance in the involved organs and usually affects people between the ages of 25 and 40. African Americans are more commonly affected in the United States, with incidence in males: females being 15.3 versus 21.6 per 100,000 population per year.[1] Lifetime incidence is around 0.8%–2.4%.[2] Some studies have shown a negative association between cigarette smoking and sarcoidosis.[3,4]

Sarcoidosis has a variety of clinical phenotypes and mainly affects the lungs and lymphatic system. Around

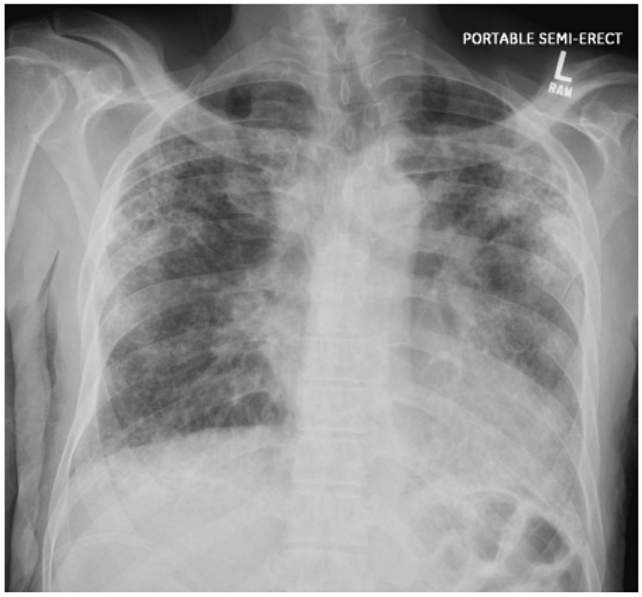

FIGURE 1 A chest radiograph showed bilateral hilar adenopathy with a reticular consolidation pattern suggestive of interstitial lung disease.

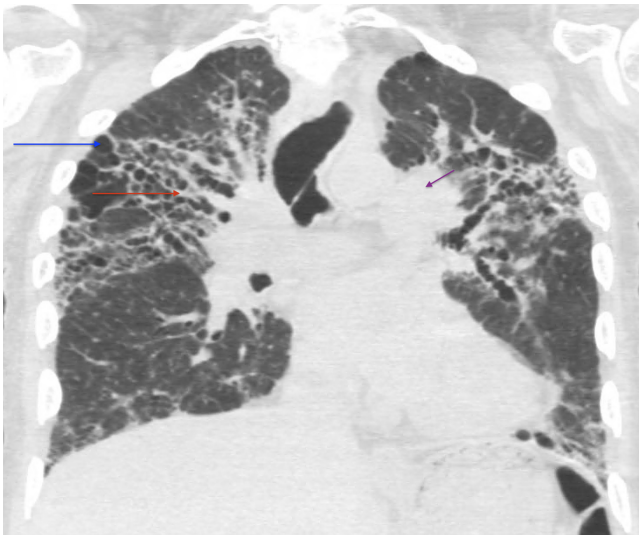

FIGURE 2 High-resolution computed tomography of the chest showing severe traction bronchiectasis of the upper lobes (red arrow) with multiple cysts in the honeycomb pattern (blue arrow), central and peripheral nodules, and mediastinal lymphadenopathy (purple arrow).

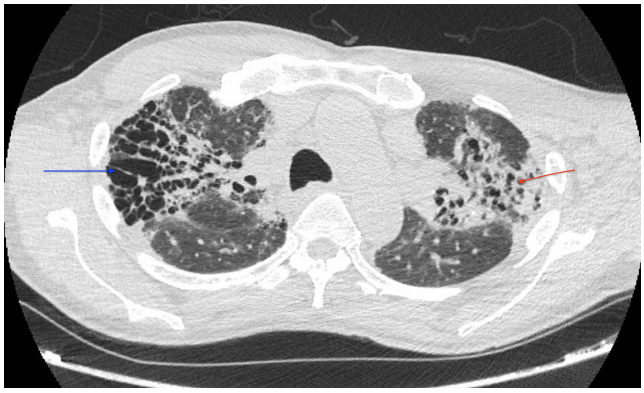

FIGURE 3 High-resolution computed tomography craniocaudal view of the chest showing severe traction bronchiectasis of the upper lobes (red arrow) with multiple cysts in the honeycomb pattern (blue arrow).

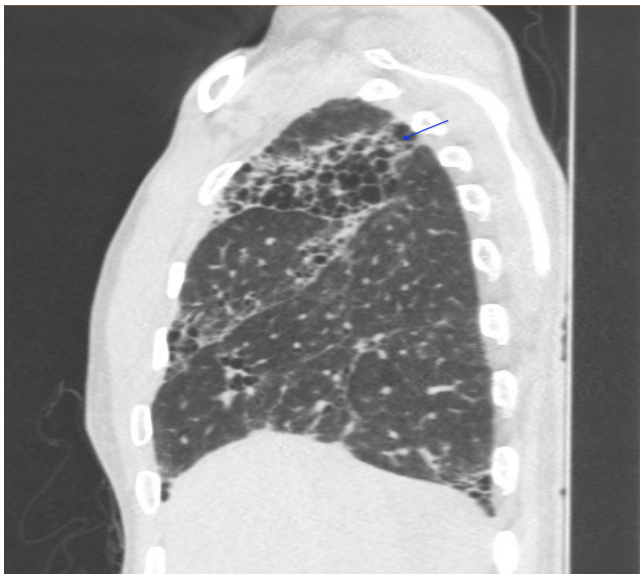

FIGURE 4 High-resolution computed tomography sagittal view of the chest showing severe traction bronchiectasis of the upper lobes (blue arrow).

95% of the patients with sarcoidosis show pulmonary findings during the course of their lives.[5] Investigations determine bilateral hilar lymphadenopathy accompanied by paratracheal lymphadenopathy in most cases. Atypical presentations like those seen in our patient pose a diagnostic challenge as clinical features and imaging findings mimic malignancy. The natural history of sarcoidosis spans from spontaneous resolution to advanced fibrocystic disease, and most of the cases have a propensity to take a benign course. As the disease progresses, traction bronchiectasis and airway angulation become evident and are reported in 40% of cases with fibrotic stages of Sarcoidosis Stage IV.[6,7] While traction bronchiectasis is predominantly seen, honeycomb lung-like cystic structures in pulmonary sarcoidosis like those seen in our patient are a distinctive process and represent the advanced stage of sarcoidosis. Pulmonary hypertension is also recognized as a complication of sarcoidosis, with a prevalence of 1%–28% of all patients at rest and up to 43% with exercise.[8–10]

The pathogenesis is supposed to be due to genetic susceptibility and environmental agents, which include possible infections like mycobacteria and propionibacteria. A strong correlation between the occurrence of sarcoidosis and human leukocyte antigens and other

polymorphisms is also seen. Interleukin (IL)-12 and IL-18, γ, and tumor necrosis factor (TNF)-α play a crucial role in the course and progression of the granulomatous process found in sarcoidosis. 86%–92% of cases show lung involvement as per chest X-ray findings.[2] The presence of a patchy focal increase in lung density with central crowding of bronchi and vessels on high-resolution computed tomography (HRCT) inclines toward the diagnosis of sarcoidosis.[11] And those with bronchiectasis findings on HRCT show a higher incidence of acute worsening events and respiratory failure. Pulmonary sarcoidosis with atypical imaging features is seen in 15%–25% of patients, and they may include diffuse ground glass opacities, multiple nodules, necrotizing consolidations, and honeycombing.[12] Histology is the gold standard for the diagnosis of sarcoidosis, which should show non-caseating granulomas with supporting evidence of disease clinically and radiographically. Serum ACE is increased in 30%–80% of cases of sarcoidosis. The occurrence of bronchiectasis is related to the mechanical stretch of the airways as a result of parenchyma fibrosis, which seldom causes bronchiectatic symptoms.

Most patients with sarcoidosis do not require treatment. Corticosteroids are the mainstay of treatment to manage unfavorable clinical courses and evolution. They are known to reverse the granulomatous process. A commonly used second-line agent is methotrexate, which helps to decrease the dose of prednisone when used with steroids. Current and past systemic sarcoidosis medications were categorized as glucocorticoids, cytotoxic drugs such as methotrexate, leflunomide, azathioprine, and anti-TNF monoclonal antibodies, or etanercept.[13,14] According to some studies, no significant difference is found in pulmonary function or the rate of occurrence of bronchiectasis among those who are receiving vs. not receiving anti-TNF antibodies. Double lung transplantation is the treatment of choice for clinical bronchiectasis, taking into consideration the risk of infection from the residual native side.[15,16] In one study, 75% of patients with Stage IV sarcoidosis died as a result of pulmonary complications, and pulmonary hypertension is directly responsible for mortality in 31.2% of cases and chronic respiratory failure in 25% of cases.[17] Respiratory failure is the most common cause of mortality due to sarcoidosis in the USA and Europe, while it is cardiac sarcoidosis in Japan.[18]

3 | CONCLUSION

In conclusion, we describe an uncommon presentation of sarcoidosis with honeycombing and cystic bronchiectasis. Nonetheless, with the possibility of active malignancy, a high index of suspicion and careful review of history and radiological imaging are necessary for timely diagnosis and optimal management. With this case report, we would like to help clinicians and researchers expand their understanding of the condition, different radiological patterns of sarcoidosis, and treatment outcomes in order to help improve the overall quality of life of patients.

AUTHOR CONTRIBUTIONS

Raja Shekar Jadav: Conceptualization; data curation; supervision; validation; visualization; writing – original draft; writing – review and editing. **Sriharsha Dadana:** Conceptualization; data curation; validation; visualization; writing – original draft; writing – review and editing. **Akshay Avula:** Supervision; validation; visualization.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

Authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

All data underlying the results are available as part of the article, and no additional source data is required.

ETHICS STATEMENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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