




An Interesting Case of Bilateral Hilar Lymphadenopathy Presented with Visual Impairment

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Abstract

Sarcoidosis is a systemic granulomatous disease of unknown etiology. It primarily affects the lungs and lymphatics; however, the presentation is not always pulmonary. Uveitis is the presenting symptom in 5% of patients, predominantly in females. Cystoid macular edema is the most important sight-threatening sequela of ocular sarcoidosis. Histopathological evidence of noncaseating epithelioid granulomas from a biopsy is the gold standard for the diagnosis of ocular sarcoidosis. Diagnosis of sarcoidosis is made by exclusion of other causes of granulomatous disease, mainly tuberculosis and fungal infection and documentation of involvement of at least one additional organ system. The authors present a case of a patient with sarcoidosis involving lungs and eyes, who had uveitis as an initial presentation.

Keywords

- ▶ EBUS TBNA
- ▶ granulomatous lesion
- ▶ sarcoidosis
- ▶ uveitis

Introduction

Sarcoidosis is a multisystem chronic inflammatory disorder of unknown etiology, characterized pathologically by noncaseating granulomas. It usually affects young adults with variable clinical presentation. The lung is the more frequently affected organ and accounts for morbidity and mortality associated with this disease. Other organs commonly involved include the eyes, skin, and lymph nodes.

The incidence and prevalence of sarcoidosis vary substantially, depending on the region of the world. It commonly

affects women than men. Diagnosis of sarcoidosis relies on compatible clinicoradiological findings and is supported by histopathological evidence of noncaseating epithelioid granulomas in involved organs, exclusion of other causes of granulomatous disease, mainly tuberculosis and fungal infection, and documentation of involvement of at least one additional organ system.

Corticosteroids are the mainstay treatment for sarcoidosis. Additional immunosuppressive agents including methotrexate, azathioprine, mycophenolate mofetil, infliximab, and adalimumab are used in refractory systemic sarcoidosis and lung transplantation in advanced lung disease.

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Case Report

A 59-year-old female presented with redness and blurring of vision in both eyes for 20 days. An ophthalmic evaluation showed panuveitis in both eyes. Physical examination and laboratory results were unremarkable. A chest X-ray was taken, which showed bilateral upper and midzone nodular opacities and hilar lymph nodes. Further contrast-enhanced computed tomography (CECT) thorax showed bilateral symmetrical parenchymal nodular and confluent peribronchovascular opacities, perilymphatic nodules, nodular thickening of the fissure, septa with upper lobe predominance, and bilateral hilar and mediastinal enlarged lymph nodes (►Figs. 1 and 2).

Flexible bronchoscopy (FB) followed by endobronchial ultrasound (EBUS) was planned and performed under moderate sedation, and no endobronchial lesions were visualized. Bronchoalveolar lavage (BAL) was taken and sent for bacterial and fungal culture. EBUS showed bilateral hilar, subcarinal, and right paratracheal nodes. EBUS-transbronchial needle aspiration (TBNA) and rapid on-site evaluation (ROSE) were done from the subcarinal lymph node and sent for cytology and GeneXpert (►Fig. 3). Mycobacterium tuberculosis was not detected in GeneXpert, and cytological examination of lymph node aspirate showed noncaseating granulomatous lymphadenitis (►Figs. 4 and 5). The BAL

culture showed no growth for both bacteria and fungus. The serum angiotensin-converting enzyme (ACE) and calcium levels were normal. In the Mantoux test, no significant induration was seen. Given the diagnosis of sarcoidosis



Fig. 1 Computed tomographic image of the chest demonstrating hilar and subcarinal lymph nodes.



Fig. 2 Computed tomographic image of the chest demonstrating numerous perilymphatic nodules and peribronchovascular opacities.

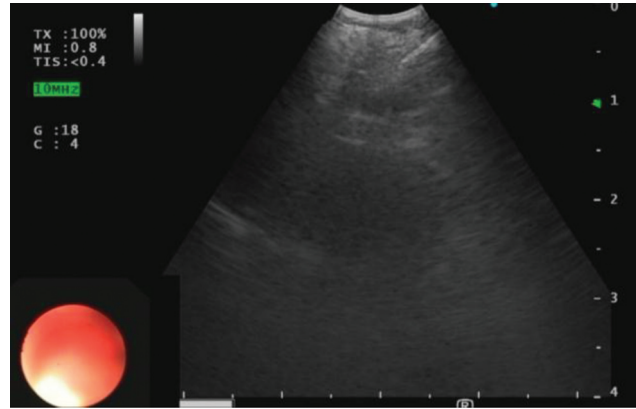


Fig. 3 Endobronchial ultrasound image demonstrating transbronchial needle aspiration from subcarinal lymph node.

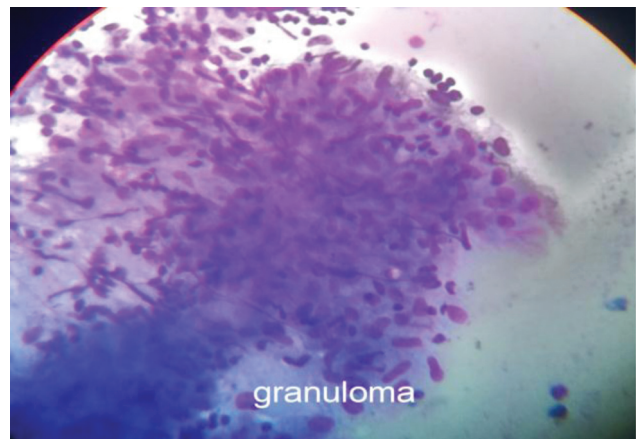


Fig. 4 Cytology of lymph node aspirate showing noncaseating granuloma.



Fig. 5 Cytology of sarcoid granuloma showing Langerhans giant cell.

above, the patient was started on methylprednisolone pulse therapy, followed by oral corticosteroids.

Discussion

Sarcoidosis is a systemic chronic inflammatory granulomatous disease of unknown etiology. It usually affects young adults with variable clinical presentation. It can affect multiple organs to a varying extent and degree; lung involvement is frequent, but the presentation is not usually pulmonary. In 20 to 50% of patients, uveitis is an early feature of sarcoidosis.¹

The incidence and prevalence of sarcoidosis vary substantially, depending on the region of the world; women are commonly affected than men. Its incidence and prevalence are estimated at 15.3 to 21.7/100,000 and 10 to 20/100,000 population, respectively.² In India, sarcoidosis is an underdiagnosed disease.

Many patients with pulmonary sarcoidosis are asymptomatic with incidental chest radiograph findings. Common presenting respiratory symptoms include shortness of breath, cough, and chest pain. Wheezing may be present in patients with endobronchial involvement. Symmetrical and bilateral hilar lymphadenopathy is a classical chest radiograph finding, and right hilar adenopathy may be slightly more prominent.³ The lung parenchymal findings on the chest radiograph include diffuse reticular or ground-glass opacities, nodular opacities, and fibrocystic changes with mid-to-upper zone predominance. Bronchoscopic findings of sarcoidosis include nodules, plaques, erythema, and cobblestone appearance.⁴

Ocular sarcoidosis accounts for 10 to 50% of cases, including lacrimal gland enlargement.⁵ In A Case-Control Etiologic Study of Sarcoidosis (ACCESS study), a prospective study, 11.8% of patients had ocular manifestations as presenting symptoms.⁶ Sarcoidosis is the leading cause of uveitis in elderly patients.⁷ In retrospective series of histologically proven sarcoidosis, symptomatic uveitis accounts for 20 to 50%.⁸ Sarcoid uveitis is typically bilateral (80–90%), with anterior uveitis being the most common, accounting for 41 to 75% of sarcoid uveitis, followed by posterior, intermediate uveitis, and panuveitis.⁹ Panuveitis accounts for 7 to 14% of sarcoid uveitis and is one of the most important systemic associations in Japan and Europe.^{10,11}

Typical sarcoid uveitis presents with mutton-fat keratic precipitates, iris nodules (Koeppe, Busacca nodules), anterior and posterior synechiae, band keratopathy, and glaucoma. Posterior involvement includes vitritis, vasculitis, and chorioidal lesions. Cystoid macular edema is the most important and sight-threatening consequence of sarcoid uveitis.

No treatment is required for asymptomatic patients. In symptomatic patients, corticosteroids are the mainstay of treatment. Additional immunosuppressive agents including methotrexate, azathioprine, mycophenolate mofetil, infliximab, and adalimumab are used in refractory systemic sarcoidosis and lung transplantation in advanced lung disease.

EBUS-TBNA with ROSE has a good diagnostic yield and assists in safe, adequate specimen sampling with rapid results. EBUS-TBNA with or without ROSE is superior to conventional TBNA (c-TBNA) without ROSE in the diagnosis of sarcoidosis.¹²

Conclusion

Uveitis is a common extrapulmonary manifestation of sarcoidosis, particularly in females; however, due to lack of investigational modalities, it is underdiagnosed in India. EBUS-TBNA is exceptionally useful in the evaluation of mediastinal lymphadenopathy and early diagnosis of diseases like sarcoidosis.

Funding

None.

Conflict of Interest

None declared.

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