Case Report

Sarcoidosis Presenting with Tracheobronchial Calcification and Nodularity: An Unusual Case Presentation with Treatment Response Assessment by ^{18}F-FDG-PET/CT

Abstract

Airway involvement, tracheobronchial nodularity, and calcification are rare occurrences and unorthodox phenomena in sarcoidosis. Here, we report such an unusual case manifesting as tracheal calcification and nodules of the central airways. Radiology and bronchoscopy provide useful diagnostic clues when combined with histopathology. This case serves as an aide-memorie for the variegated presentations of sarcoidosis and emphasizes a high index of suspicion for the entity in such unconventional circumstances. An early favorable treatment response assessment to corticosteroid therapy was shown with ^{18}F-fludeoxyglucose positron emission tomography/computed tomography.

Keywords: *Bronchoscopy, PET-CT, tracheal calcification*

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Introduction

Sarcoidosis is a multisystem disease characterized by noncaseating granulomas most commonly involving the lung parenchyma and mediastinal lymph nodes. Two-third of the cases can have airway involvement, though it is rarely seen in isolation. The entire extent of the airways from the nasal passage up to terminal bronchioles can be affected. Here, we present a rare case of predominant tracheobronchial involvement by sarcoidosis manifesting as tracheal calcification radiologically, mucosal nodules bronchoscopically and with high metabolic uptake on ^{18}F-fludeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG-PET/CT). This unconventional presentation of sarcoidosis has been sporadically reported in the literature.

Case Report

A 49-year-old man was referred with a history of dry cough and exertional breathlessness of 2 months duration. Medical history was significant for type II diabetes mellitus (DM) treated with oral hypoglycemic agents. General examination revealed tachypnea. Respiratory system examination revealed bilateral rhonchi. Hemotological and biochemical blood parameters were normal. The chest X-ray was normal. CT of the thorax [Figure 1] showed circumferential nodularity, calcification of the trachea-bronchial tree, and patchy consolidation in bilateral lower lobe areas. The differentials of tracheopathica osteochondroplastica (TPO), tuberculosis, sarcoidosis, and vasculitis were considered. Sputum evaluation was negative for acid fast bacilli (AFB). Spirometry was suggestive of an obstructive abnormality, with forced

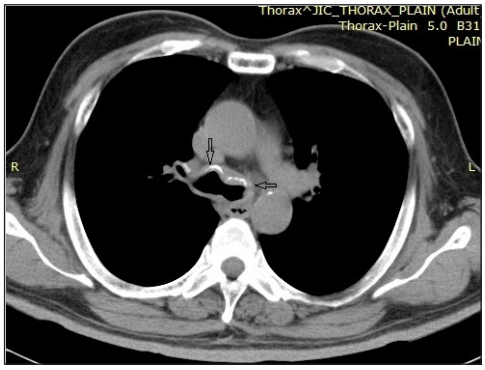

Figure 1: Computed tomography of thorax showing circumferential calcification of the trachea-bronchial tree, and patchy consolidation in bilateral lower lobe areas

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Access this article online

Website: www.ijnm.in

DOI: 10.4103/ijnm.IJNM_152_16

Quick Response Code:

How to cite this article: Bansal S, Utpat K, Desai U, Basu S, Joshi JM. Sarcoidosis presenting with tracheobronchial calcification and nodularity: An unusual case presentation with treatment response assessment by ^{18}F-FDG-PET/CT. Indian J Nucl Med 2017;32:217-20.

inspiratory volume in 1st second ($FEV_{1)}$ of 0.75 L (35% of predicted) with good bronchodilator reversibility. Fiberoptic bronchoscopy (FOB) demonstrated widespread nodularity of the entire trachea-bronchial tree [Figure 2], extending to the segmental bronchi. Endobronchial biopsy (EBB) of the nodules revealed noncaseating granulomas on histopathology. EBB AFB culture was negative. Bronchial washings and post-scopy sputum were evaluated for GeneXpert, which did not detect *Mycobacterium tuberculosis* (MTB). Mantoux test was negative. Serum ACE level was 32 IU/ml. Perinuclear-Anti-Neutrophil-Cytoplasmic-Antibodies (P-ANCA) and Cytoplasmic-Anti-Neutrophil-Cytoplasmic-Antibodies (C-ANCA) were negative. A provisional diagnosis of sarcoidosis was kept. ^{18}F-FDG-PET/CT [Figure 3] was performed, which showed enhanced metabolic activity in the nodular circumferential wall thickening with calcification in the trachea, extending distally up to the main bronchi and proximal lobar bronchi. Summary of results has been provided in Table 1. The patient was treated with oral prednisolone (40 mg daily) with inhaled corticosteroids (ICS) and long-acting beta-2 agonist (LABA) combination administered through a pressurized metered dose inhaler (MDI). On 6-week follow up, the patient had complete resolution of symptoms, improvement in FEV_1 by1500 ml, complete resolution of nodularity on bronchoscopy [Figure 3]; repeat ^{18}F-FDG-PET/CT [Figure 4] demonstrated resolution of hypermetabolism observed in the baseline scan, suggesting favorable metabolic response to therapy.

Table 1: Summary of investigations

Serology	Reports
ACE	32 IU/ml (WNL)
p-ANCA/c-ANCA	Negative
Microbiology	
Sputum AFB Smear	Negative
Sputum GXP	MTB not detected
Bronchial Washing GXP	MTB not detected
Radiology	
CT Thorax	Circumferential nodularity, calcification of the trachea-bronchial tree, and patchy consolidation in bilateral lower lobe areas.
PET-CT	Enhanced metabolic activity in the nodular circumferential wall thickening with calcification in the trachea, extending distally up to the main bronchi and proximal lobar bronchi. Focal uptake in the left lower lobe.
Spirometry	Obstructive abnormality with good reversibility.
Histopathology	Noncaseating granulomas

Discussion

Tracheal affection can be seen in a number of primary disorders and systemic diseases. The trachea is sometimes referred to as the "forgotten zone" as the pathological processes involving trachea may not receive prominent clinical consideration in disorders presenting with respiratory symptoms and signs.[1] Multidetector computed tomography (MDCT), the imaging of choice, provides high-resolution images with multiplanar reformations, minimum intensity projections, and three-dimensional and virtual bronchoscopic images.[2] Tracheal calcification has a variety of differential diagnosis such as relapsing polychondritis (RP), TPO, amyloidosis, sarcoidosis, and Wegner's granulomatosis, among others. RP and TPO have hard gritty nodules that spare the posterior tracheal

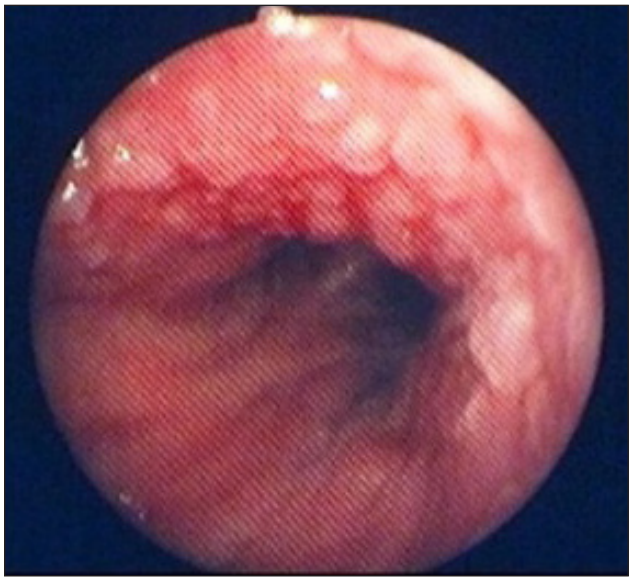

Figure 2: Fiberoptic bronchoscopy showing widespread nodularity of the entire trachea-bronchial tree extending to the segmental bronchi

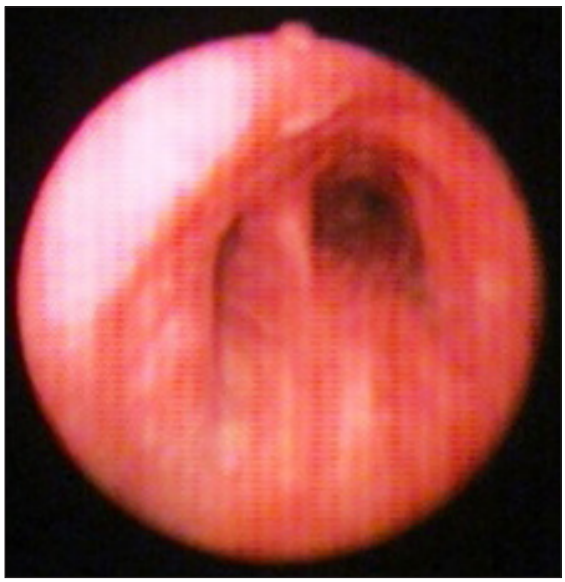

Figure 3: Repeat fiberoptic bronchoscopy at 6 weeks showing total resolution of the nodularity of the entire trachea-bronchial tree

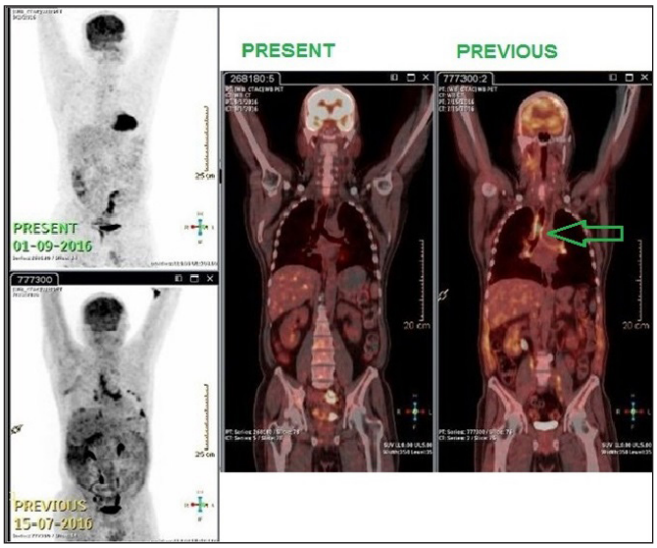

Figure 4: Comparative images of previous (baseline) and present (at 6 weeks follow up) ^{18}F-FDG-PET/CT scans. The baseline scans show enhanced metabolic activity in the nodular circumferential wall thickening with calcification in the trachea, extending distally up to the main bronchi and proximal lobar bronchi (green arrow)

wall unlike in our case.[3] Amyloidosis and vasculitis have distinct histopathological features and have associated systemic involvement. Sarcoidosis is a heterogeneous multisystemic chronic inflammatory condition characterized by noncaseous epithelioid cell granulomas, which affect almost any organ. Lung, mediastinal, and hilar lymph nodes are most commonly involved in 90% of the cases.[4] Airway involvement, as judged by clinical features, physiologic testing, imaging techniques, bronchoscopy, and biopsy is observed in two-thirds of patients.[5] It is divided as sarcoidosis of the upper respiratory tract (SURT) involving structures from the nares to vocal cords and sarcoidosis of the lower respiratory tract involving the trachea, the bronchial tree, and small airways. Isolated tracheobronchial involvement without any parenchymal affection is uncommon.[6] Air-trapping in sarcoidosis correlates with small airways disease is reported in 20% of the patients. The resultant cough and wheezing prompt patients to seek medical help, which is confirmed on spirometry.[7] Our patient had excessive dry cough and wheeze and rhonchi on auscultation.

FOB has a noteworthy role in visualization and tissue sampling. The classical endobronchial sarcoidosis manifests as mucosal islands of waxy yellow mucosal nodules, 2-4 mm in diameter, nonuniform with increased profusion seen towards the lobar, and segmental bronchi as compared to the central airways.[8] Occasionally, nodules may coalesce leading to a cobblestone appearance. Sarcoid granulomas culminating in endobronchial obstruction may simulate an obstructing malignant mass.[9] Our patient had similar trachea-bronchial nodularity but with a diffuse distribution. Various FOB-guided biopsy options include transbronchial lung biopsy (TBLB), EBB, transbronchial needle aspiration (TBNA) of the mediastinal lymph nodes.[10] In our case, EBB of nodules confirmed sarcoidosis on histopathology and ruled out tuberculosis on microbiology.

^{18}F-FDG-PET/CT, in recent years, has demonstrated substantial promise in systemic inflammatory disorders in defining the extent of the disease, target a suitable location for biopsy, and uncover extrathoracic affection; additionally, the baseline scan works as the basis for early monitoring of therapeutic efficacy of the administered treatment.[11-13] Although not indicated in the standard workup, it can be of great value to complement the more conventionally used techniques.

Conclusion

To summarize, sarcoidosis should be considered as a differential in predominant airway involvement, especially with tracheal calcification. Newer techniques such as ^{18}F-FDG-PET/CT coupled with FOB aid in early diagnosis. Dearth of knowledge about this uncommon presentation can lead to a poor vigilance for the phenomenon, delayed diagnosis, irrevocable complications, and undue morbidity. Treatment is individually tailored taking into consideration the clinical picture, disease activity, and lung function limitation.

Financial support and sponsorship

Nil.

Conflicts of Interest

The authors declare no conflicts of interest.

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