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Case Report

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Diffuse Panbronchiolitis - Report of a Rare Case with Review of Literature

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ABSTRACT

A case of diffuse Panbronchiolitis in a 71-year-old female is presented here, because of its rarity in the Indian population. It was mostly reported from East Asian countries. However, to date, only very few cases have been reported in the literature from the Indian subcontinent. It might be due to low prevalence among different ethnical backgrounds of the Indian population, or due to underreporting in India because of a lack of awareness and recognition.

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Introduction

Diffuse Panbronchiolitis (DPB) is an inflammatory lung disease, and was first identified in 1969. It was mostly reported from East Asian countries such as Japan, China, and Korea [1]. The name, Diffuse Panbronchiolitis, was proposed to distinguish it from chronic bronchiolitis. "Diffuse" refers to "presence of lesions in both the lungs," and "pan" refers to "inflammation in all layers of bronchioles." In the years following the initial description of the DPB in Japan, cases were also identified in other parts of Asia, including China, India, and Taiwan. Over the years, DPB cases have been reported from across the globe. To the best of our knowledge, only three proven cases have been reported from India till now [2]. Different ethnic backgrounds of the Indian population (Caucasians, not Asians) might be one reason for low DPB reporting in the Indian subcontinent, despite being part of Asia. However, the effects of DPB underreporting in India due to a lack of awareness and recognition, leading to misdiagnosis and delayed treatment, cannot be ignored. The prognosis of DPB has significantly improved after the introduction of long-term, low-dose macrolide therapy in 1985 [3].

Case Report

A 71-year-old female, known case of chronic rhinosinusitis with a history of seronegative rheumatoid arthritis and recurring lower respiratory tract infections for 30 years, presented to the emergency department (ED) with worsening shortness of breath and cough with scanty sputum. At presentation, she was in respiratory failure. She reported a significant worsening of her respiratory symptoms in the last 3 years, requiring oxygen support during exacerbations. She had taken multiple courses of oral, IV, and inhaled steroids throughout the course of her disease. She also gave a history of various microbial growths isolated in sputum cultures, including Pseudomonas and H. influenzae, through the course of her recurring illness. X-ray Chest showed hyperinflation and increased bronchial markings (Figure 1A). Her HRCT thorax showed multiple centrilobular nodules with tree-in-bud opacities and areas of bronchiolectasis with bronchiolar wall thickening distributed bilaterally, more in the basal segments of the bilateral lower lobe, and also patchy bronchiolectasis bilaterally (Figure 1B, Figure 2, Figure 3, Figure 4). She underwent a fibreoptic bronchoscopy for sampling. Sputum culture, Sputum acid-fast bacilli, fungal culture, and TB-PCR were all negative. She also complained of a persistent headache for which nasal endoscopy was performed, and a diagnosis of sinusitis was made. Considering her chronic progressive symptoms leading to respiratory failure, chronic sinusitis, consistent radiographic findings, and failure to respond to multiple courses of antibiotics and pulse steroids, a diagnosis of diffuse Panbronchiolitis was entertained, and she was initiated on oral azithromycin with regular outpatient follow-up. A cold agglutinin test was also done, which showed positivity. After 3 months of macrolide therapy, the patient's respiratory symptoms significantly improved with no further steroid or antibiotic courses. CT imaging showed near resolution of the bronchiolitis and tree-in-bud nodularity, and pulmonary function testing showed a dramatic improvement in her obstructive ventilatory defect.

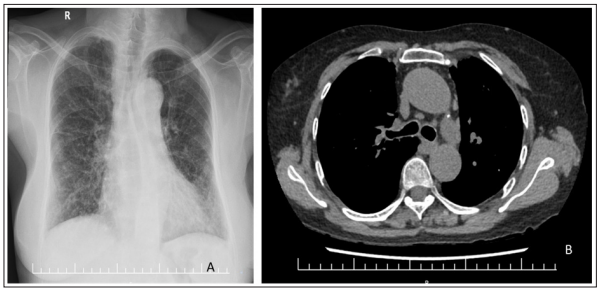

Figure 1A: X-ray Chest PA View Showing Hyperinflation and Increased Bronchial Markings Figure
1B: CT Thorax Mediastinal Window

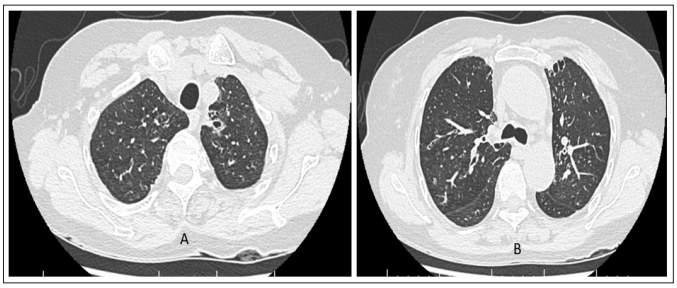

Figure 2 A & B: Minimal Bronchiectasis in the Left Upper Lobe

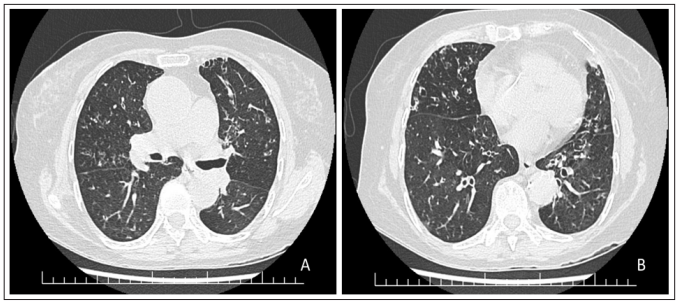

Figure 3 A & B: Bronchiectatic Changes, Nodular Shadows, and Linear Shadows Suggestive of Bronchiolar Involvement

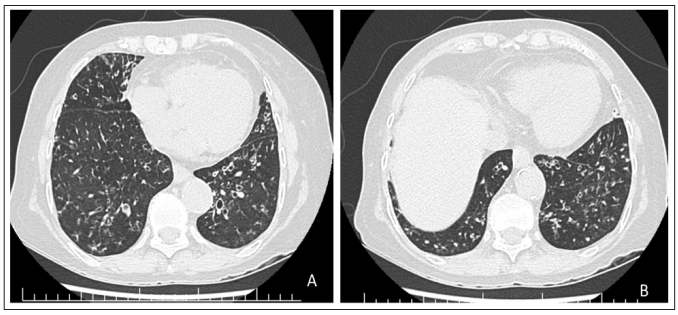

Figure 4 A & B: Bilateral Nodular Lesions and Linear Shadows Suggestive of Bronchiolectasis. Dilated Bronchi are also Seen

Discussion

Diffuse Panbronchiolitis (DPB) is an idiopathic inflammatory disease characterized by bronchiolitis and sinusitis. This entity principally effects the respiratory bronchioles, causing progressive suppurative and severe obstructive respiratory disorder [4]. Untreated DPB can lead to respiratory failure within a few years, with nearly 50 percent of untreated patients dying within five years of diagnosis [5]. Symptoms are often progressive and mimic common respiratory illnesses like bronchitis and bronchiolitis, and the diagnosis is made through a combination of clinical symptoms, imaging, and occasionally histopathology [6]. There are no specific laboratory or radiographic abnormalities that are diagnostic for DPB.

Diagnosis is made through characteristic findings that raise the suspicion for DPB, including recurrent H. influenzae or Pseudomonas aeruginosa infections, an obstructive ventilatory defect, and diffuse tree-in-bud nodularity. The clinical profile typically involves patients in the 2nd to 5th decades of life with a history of chronic paranasal sinusitis. The typical order of symptom onset includes cough with sputum, followed by the onset of exertional dyspnoea. Physical examination may reveal crackles or wheeze or both [4]. Plain chest radiography may reveal bilateral, diffuse, small nodular shadows with pulmonary hyperinflation. In advanced cases, ring-shaped or tram-line shadows can be observed, which may suggest bronchiectasis [4]. HRCT scans (Figure 5) often show a "tree-in-bud" pattern, bronchiolar wall thickening, and bronchiectasis [7,8].

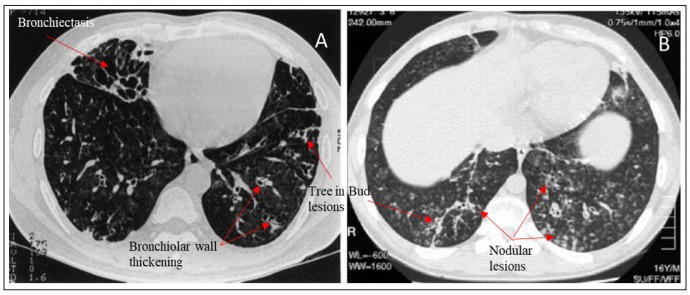

Figure 5: HRCT Findings in DPB Characterised by Tree in Bud Opacities, Bronchiolar Wall Thickening and Bronchiolectasis

A characteristic laboratory feature is persistent elevation of cold agglutinins with tests for Mycoplasma pneumoniae negative test for Mycoplasma [4]. Markedly high levels of the "tumour-associated" carbohydrate antigens sialyl stage-specific embryonic antigen-1 and sialyl Lewis (a) have been demonstrated in the serum and BALF of patients with DPB [8]. Pulmonary function tests show a significant airflow limitation, which is relatively resistant to bronchodilators [9]. Analysis of arterial blood gas usually shows hypoxaemia (partial pressure of arterial oxygen <80 mmHg). The diffusing capacity is variably reduced. In advanced DPB, P. aeruginosa is superinfected, reducing the lungs' capacity for gas exchange, which brings about the progression of hypoxaemia and, later, hypercapnia. This eventually results in the development of Pulmonary hypertension, leading to cor pulmonale and consequent death due to chronic respiratory failure in most cases [4].

The histopathologic diagnostic criteria include severe inflammation in all layers of the respiratory bronchioles and lung tissue lesions that appear as nodules within the terminal and respiratory bronchioles in both lungs [4].

A significant improvement in the prognosis of this potentially fatal disease has been reported with the use of long-term therapy with macrolide antibiotics, the effect of which is attributed to anti-inflammatory and immunoregulatory action Because appropriate macrolide therapy results in a dramatic improvement in prognosis (>90% 10-year survival rate), a clinician should consider the diagnosis of DPB despite its rarity and nonspecific presentation [4]. In certain circumstances, obtaining a biopsy may not be feasible, and modalities such as HRCT thorax and response to treatment may be considered to make an appropriate diagnosis.

In this case, a diagnosis of seronegative rheumatoid arthritis (RA) was made earlier. RA can lead to respiratory bronchiolitis and bronchiolectasis. So, RA-related chronic bronchiolitis is a close differential diagnosis. However, repeated serology failed to establish any collagen vascular disease, and despite a diagnosis of RA for 30 years, she did not develop any joint deformity to prove the diagnosis.

Conclusion

We report a rare case of diffuse Panbronchiolitis based on the constellation of symptoms, disease progression, and HRCT findings. The patient had multiple courses of steroids and antibiotics before ultimately being diagnosed with DPB. One should entertain the diagnosis of DPB in appropriate clinical settings despite its rarity and nonspecific presentation. In certain circumstances, obtaining a biopsy may not be feasible, and

modalities such as HRCT thorax and response to treatment may be considered to come to an appropriate diagnosis. The advent of macrolide therapy has changed the prognosis and clinical outcome of the disease to a remarkable extent and hence, coming to an accurate diagnosis can have a significant impact on the quality of life and prognosis of patient.

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