Open Journal of
Clinical & Medical
Case Reports

Volume 8 (2022)
Issue 21

ISSN: 2379-1039

Pleuro-parenchymal fibroelastosis with combined pulmonary fibrosis with emphysema: A case report

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Abstract

Background: This report highlights the first published case of Pleuroparenchymal Fibroelastosis (PPFE) associated with combined pulmonary fibrosis with emphysema.

Case presentation: We report a case of above association of a 67 year old patient who has presented with MRC grade 2 dyspnea with anorexia without fever or hemoptysis. He has been treated as smear negative Pulmonary Tuberculosis 36 years back. He was emaciated and had deepened supraglottic notch. In addition, he had loud pulmonary component of the second heart sound.

High resolution CT (HRCT) showed bilateral upper zone pleural thickening with adjacent fibrotic changes and para-septal emphysema in the upper lobes with lower zone predominant honey combing with traction bronchiectasis involving extreme bases. 6 minute walking test showed significant desaturation from 97% to 90% with a walking distance of 1100 feet.

Spirometry showed combined restrictive and obstructive pattern (FVC – 52% , FEV1- 48%, FEV1/FVC – 61%). Bronchial wash did not show any evidence of infection including a negative Gene X-pert. 2D echo showed mild pulmonary hypertension (TRPG -41 mmHg) with normal cardiac function. HRCT was suggestive of PPFE, and combined pulmonary fibrosis with emphysema. He was started on Tiotropium inhalers and Pirfenidone with close follow up.

Conclusion: This case highlights a rare interstitial lung disease associated with another interstitial lung disease which has never been described in literature. Clinical examination and detailed radiological evaluation supports the diagnosis. PPFE is commonly associated with an aetiological agent in most circumstances.

Keywords

Emphysema; Interstitial lung disease; Pleuro-parenchymal fibroelastosis; UIP.

Abbreviations

CPFE: Combine Pulmonary Fibrosis and Emphysema; CXR: Chest X-ray; FEV1: Forced Expiratory Volume in 1st second; FVC: Forced Vital Capacity; HRCT: High Resolution Computed Tomography; ILD: Interstitial Lung Diseases; NSIP: Non Specific Interstitial Pneumonia; PPFE: Pleuro Parenchymal Fibroelastosis; TB: Tuberculosis; TRPG: Tricuspid Pressure Gradient; UIP: Usual Interstitial Pneumonia.

Introduction

Pleuroparenchymal Fibroelastosis (PPFE) is a rare Interstitial Lung Disease (ILD) with unique clinical, radiological and pathological characteristics. There are number of case reports stating its association with different other ILD. However, there are no case reports to describe its association with usual interstitial pneumonia (UIP) and paraseptal emphysema involved in the same patient making a diagnosis of combined pulmonary fibrosis with emphysema syndrome (CPFE). Here we describe the first case of PPFE complicated with CPFE and pulmonary hypertension.

Case Presentation

67 year old patient presented with exertional dyspnea for 1 week associated with anorexia and weight loss for 3 months. He did not experience cough, wheezing or chest pain. He denied having orthopnea, paroxysmal nocturnal dyspnea or ankle swelling and was treated as smear negative pulmonary TB 36 years back. He was an ex-smoker with 12 pack years.

On examination he was emaciated, had deepened supraglottic notch (Figure 1) without pallor, clubbing or lymphadenopathy. His trachea was in the midline and there was bilateral fine end inspiratory crepitation mainly in the lung bases with platythorax. There were no additional lung signs in the upper zones. In addition he had loud pulmonary component of the second heart sound.

His complete blood count and inflammatory markers were within normal limits. Chest X-ray (CXR) showed upper zone pleural thickening with fibrotic changes predominantly involving upper and lower zones. His HRCT showed bilateral upper zone pleural thickening with adjacent fibrotic changes (Figure 2). In addition, there was para-septal emphysema in the upper lobes and honey combing with traction bronchiectasis predominantly in the lower zones (Figure 3) involving extreme bases. 6 minute walking test showed significant desaturation from 97% to 90% with a walking distance of 1100 feet. Spirometry showed combined restrictive and obstructive pattern (FVC – 52%, FEV_1- 48%, FEV1/FVC – 61%. TLC – 74%, DLCO – 42%). Bronchial wash did not show any evidence of infection including a negative Gene X-pert. 2D echo showed mild pulmonary hypertension (TRPG -41 mmHg) with normal cardiac function.

A radiological diagnosis of possible UIP and paraseptal emphysema suggestive of CPFE with PPFE was made. He was started on Pirfenidone gradually increasing up to a dose of 600 mg three times a day with Tiatropium inhalers with pulmonary rehabilitation and follow up was arranged. It was noted to have gradually worsening lung function in the follow up (<10%).

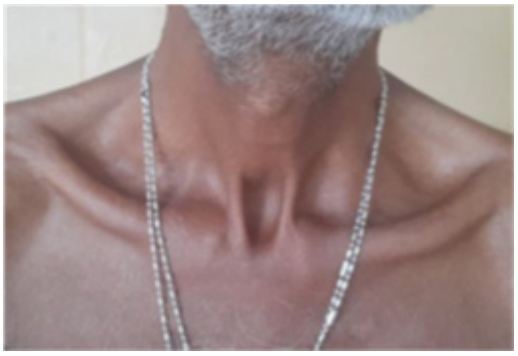

Figure 1: Deepened suprasternal notch.

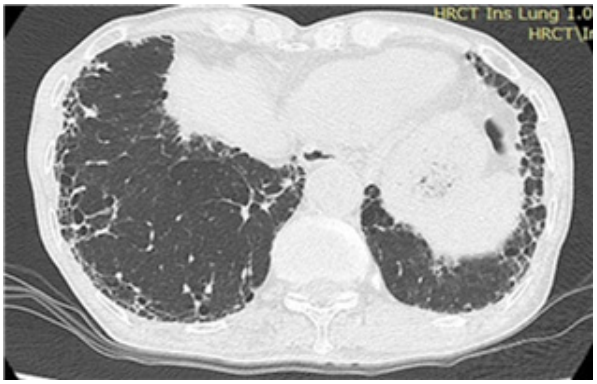

Figure 3: Honeycombing and traction bronchiectasis in the bases suggestive of UIP. There are no ground glass densities.

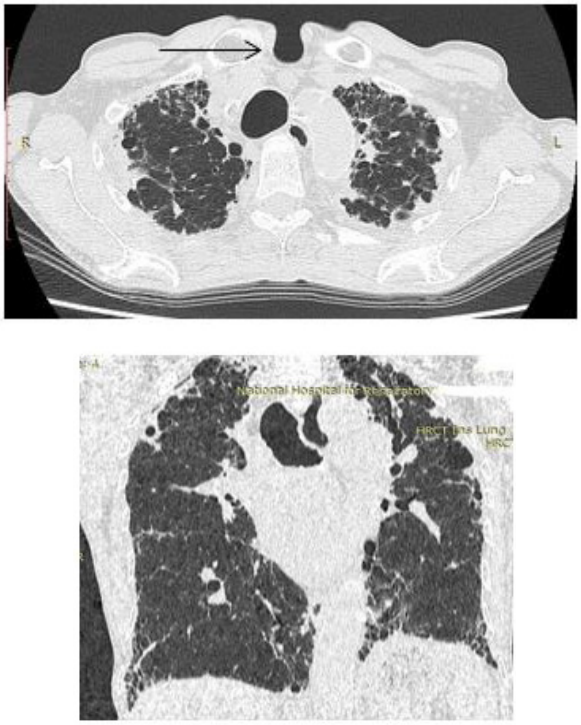

Figure 2: Bilateral upper zone pleural thickening with adjacent fibrotic changes. Posterior wall of the trachea rests on the anterior surface of the vertebra. Significant deepened supraglottic notch is also seen (arrow).

Discussion

PPFE has been described as a rare idiopathic interstitial disease in 2013 [1] although it was described at different times before. From then number of case reports and case series were described and different associations were identified including connective tissue disorders, post-transplant status [5-7] and infections. In addition it was described in association with other interstitial lung diseases such as UIP [13], Hypersensitive pneumonitis (HP) [9] etc. According to recent literature idiopathic entity is becoming less as increased numbers of causes are identified [2].

Anorexia and weight loss are common complaints and examination may reveal cachexia, deepened suprasternal notch, platythorax and upper lobe fibrotic findings. Deepened suprasternal notch led to the suspicion of the disease in our case and is a characteristic feature of PPFE. Diagnosis is usually made through radiological and histological evidence although no formal diagnostic criteria are made. However different groups have suggested certail diagnostic criteria although thre is no uniform agreement [14,15]. HRCT changes are mainly seen in the upper lobes which includes pleural thickening and adjacent parenchymal fibrosis which progresses over time. One characteristic feature that can be seen in HRCT is the resting of the posterior wall of trachea on anterior surface of the vertebrae [2] which is seen in our case as well. This feature is mostly seen on the right side due to unexplained reasons. Most of the cases are described as idiopathic although different etiologies are postulated including infections. In our case he gives a history of sputum negative TB which can be a possible etiological agent which may have initiated the injury. On the

other hand misdiagnosis of the original disease of PPFE as TB is also possible as Tuberculosis is commonly seen on our setting involving upper lobes.

Histology plays a crucial role in the diagnosis of PPFE. Our patient was not subjected to a biopsy considering his poor lung functions and the risk of complications as his disease was associated with UIP and emphysema. In addition PPFE itself is associated with a high risk of pneumothorax further increasing the burden of post biopsy complications. Apart from this, biopsy in ILD is no longer advised as it is associated with higher mortality.

It has been revealed that PPFE can get associated with other ILD including UIP, NSIP and HP. Our case become unique as CPFE has never been described in association with PPFE. On the other hand it postulates a bad prognosis as the underlying diseases are UIP [4], emphysema and PPFE which individually has a poor prognosis. As the patient had an entity of UIP he was started on Pirfenidone to reduce the disease progression.

Spirometry revleaed a FEV1 of 48% and a FEV1/FVC ratio of 61% suggesting an obstructive element and FVC of 52% showing a restrictive component as well. In addition there was an element of air trapping further convincing of obstructive component. 6MWT showed 7% desaturation from 97% of resting saturation showing a significant involvement of lung parenchy a mostly due to the fibrotic element. In this unique case it is associated with two fibrotic ILDs involving the apical area and basal area by PPFE and UIP respectively.

There is no successful treatment to PPFE and it is associated with progressive deterioration of lung function leading to death. Closer follow up with management of complications is advised and progression of the disease is extremely variable.

Conclusion

PPFE is a rare interstitial lung disease which can get associated with other ILDs. Prominent suprasternal notch is a clue to suspect PPFE in clinical examination. PPFE is associated with different disease entities and idiopathic entity is becoming less. Tuberculosis can be a possible etiology and a detailed evaluation is necessary to find out different etiological agent.

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Manuscript Information: Received: October 14, 2022; Accepted: November 10, 2022; Published: November 21, 2022

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Citation: Wickramasinghe DSA, Palihawadene S, Rathnapala A, Kularatne WNS. Pleuro-parenchymal fibroelastosis with combined pulmonary fibrosis with emphysema: A case report. Open J Clin Med Case Rep. 2022; 1935.