Indian Journal of Pathology and Oncology 2023;10(2):182–184

Content available at: https://www.ipinnovative.com/open-access-journals

Indian Journal of Pathology and Oncology

Journal homepage: www.ijpo.co.in

Case Report

Autopsy findings in a case of idiopathic pleuroparenchymal fibroelastosis: A rare entity with review of literature

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ARTICLE INFO

Article history:
Received 24-01-2023
Accepted 10-03-2023
Available online 17-06-2023

Keywords:
Idiopathic pleuroparenchymal fibroelastosis
Interstitial pneumonia

ABSTRACT

Idiopathic pleuroparenchymal fibroelastosis is a rare idiopathic interstitial pneumonia which mainly affects the upper lobe of lungs resulting in intra alveolar fibrosis, elastosis and visceral pleural fibrosis. For definite diagnosis, radiographic findings as well as histopathology findings are required. The overlap with interstitial lung diseases seen usually, makes it challenging to diagnose. There are no guidelines available for the diagnosis and appropriate management of this disease and only a few cases reported in the literature. We report a case of pleuroparenchymal fibroelastosis in autopsy in a 32-year-old female who allegedly died of heart failure with review of literature.

1. Introduction

Idiopathic upper lobe pulmonary fibrosis, also known as idiopathic pleuroparenchymal fibroelastosis (IPPFE), was initially identified in 1992 by Amitani et al. It received recognition as an uncommon form of idiopathic interstitial pneumonia with distinctive clinical, radiological, and pathological features from the American Thoracic Society and European Respiratory Society in 2013.[1] It is distinguished by pleural fibrosis and subpleural lung parenchyma that primarily affects the higher lobes. Intra-alveolar fibrosis and elastosis along with visceral pleural fibrosis are hallmark histopathological findings.[2]

Although the etiopathogenesis of this illness is still unknown, it is believed that lung or bone marrow transplants, chemotherapy, connective tissue disease, repeated pulmonary infections, and occupational dust inhalation are the causes. IPPFE must be diagnosed based on the patient's classic upper lobe predominant interstitial lung disease, pleural thickening on CT scan, and classic findings on histology.[3] Due to overlap with other interstitial lung illnesses that might result in incorrect diagnosis and unreliable detection, it may be more prevalent than what has been documented in the literature.

2. Case Report

A 32-year-old female brought dead to civil hospital, allegedly died due to heart failure as per police papers. Post mortem examination was done at civil hospital and the viscera including brain, heart, pieces of bilateral lungs, liver, spleen and bilateral kidneys were sent to the department of pathology for histopathological examination. The female was a housewife, non smoker and had no previous history of any heart or lung disease.

On gross examination, the lung pieces were heavy and congested. Cut section of lung revealed few areas of fibrosis while the other organs appeared unremarkable. Microsections examined from pieces of lung show congestion, edema, subpleural fibrosis, visceral pleural fibrosis, elastosis and perivascular fibrosis. Special stains - Mason trichrome and Van Gieson were applied

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https://doi.org/10.18231/j.ijpo.2023.037

which revealed perivascular elastosis. Keeping in view of absence of any causative factor including exposure to chemoradiation, dust exposure active infection, autoimmune disease, hypersensitive pneumonitis and other interstitial pneumonias, diagnosis of idiopathic pleuroparenchymal fibroelastosis is made. Hematoxylin-eosin stained sections from heart, brain and both pieces of kidneys show mild congestion while from liver and spleen are essentially unremarkable.

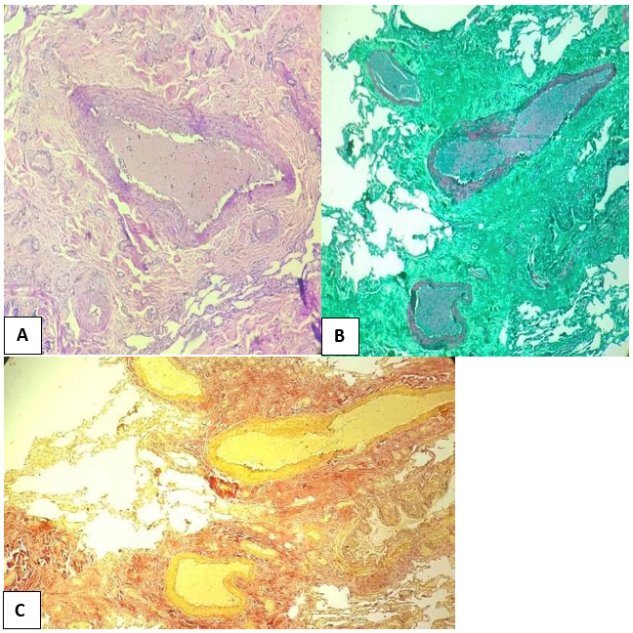

Fig. 1: A): H&E Stain, **B**): Mason trichrome stain and **C**): Van gieson stain: Section from lung showing perivascular elastosis

3. Discussion

Idiopathic interstitial pneumonia of unusual kind, or IPPFE, has an unidentified prevalence. Females with low BMI are more likely to get it, and the median survival time is only approximately 24 months. PPFE is characterised by upper-lobe predominance, pleural based parenchymal fibrosis, sparing lung bases, and peri hilar area. Although the pathophysiology is still unclear, it is thought to involve diffuse alveolar injury that causes interstitial inflammation and fibrosis as a result.[4] Lung, bone marrow, and stem cell transplantation; fibrotic interstitial lung disease; recurrent pulmonary infection; autoimmune disease; and environmental exposure to asbestos or aluminium are among the suggested triggering factors for the development of IPPFE. Additionally, a more severe manifestation of the illness has been connected to abnormalities in genes that affect telomere function.[5]

In a study done in 2020 on 10 patients, Kinoshita et al. discovered that 9 of the patients had cells that were multi-cytokeratin positive lining the inner surface of slit-like areas embedded in the zonal elastosis. Zonal elastosis as a result of alveolar collapse may be the primary lesion in IPPFE. The research established two stages of fibroelastosis development in patients with IPPFE: the early stage, when the alveolar structure collapses and forms the zonal elastosis, and the advanced stage, when intra alveolar fibroelastosis develops next to the zonal elastosis and spreads inward to form two-layered fibroelastosis.[6]

Reddy et al. performed a study on 12 patients with PPFE in 2012 with the goal of reviewing cases meeting published imaging and histological criteria and identifying any clinical features that suggest an underlying cause or pathogenesis for a condition that had previously been thought to be idiopathic. Although 25% of the individuals had coexisting UIP in the lower lobes and other instances showed symptoms of PPFE in other zones, either on HRCT or lung biopsy, review of their clinical data demonstrated that this condition seemed to be a distinct clinicopathological entity.[7]

Cheng et al. reviewed pleuroparenchymal fibroelastosis of the lung in 2016 to explore the clinical, radiological, and pathological features associated with PPFE. They also stressed that a conclusive diagnosis of PPFE necessitates a multidisciplinary approach that includes the input of a clinician, readiologist, and pathologist.[8]

Interstitial lung disease with concomitant fibrosis may be mistaken for PPFE while determining the differential diagnosis of the condition. Although a steady course is typical, incidences of rapidly worsening have been documented. The majority of PPFE patients see disease progression following diagnosis, and the prognosis for this illness is dismal. Older age, dyspnea, pneumothorax occurrences, lower body mass index (BMI), coexisting ILD, the typical pattern of interstitial pneumonia (UIP) in the lower lobes, and lower elector spinae muscle attenuation on computed tomography have all been indicated as good prognostic markers (CT). A new prognostic prediction model (PPFE Prognosis Score) for PPFE patients was proposed in a recent study by Kinoshita et al. on 104 patients. It makes use of the following variables: FVC, history of pneumothorax, ILD in the lower lobes, and serum KL-6 level.[9] Except for lung transplantation, PPFE has no effective treatment. There are only a few examples published in the literature, and there are no guidelines yet on how to treat this illness properly. To develop improved diagnostic and treatment criteria, more research is needed.

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Cite this article: Verma R, Ahalawat A, Kalra R, Tomer P, Singh S. Autopsy findings in a case of idiopathic pleuroparenchymal fibroelastosis: A rare entity with review of literature. *Indian J Pathol Oncol* 2023;10(2):182-184.