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A VERY RARE CASE OF PLEURO-PARENCHYMAL FIBROELASTOSIS (PPFE)

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ABSTRACT
Pleuro-parenchymal fibroelastosis (PPFE) is a rare ILD added to a subgroup of Idiopathic interstitial pneumonia. It is a rare disease usually involving the upper lobe. It has been found to affect the visceral pleura causing fibrosis and adjacent lung parenchyma causing fibroelastosis of the lining of the lung. Till date only 120 cases have been reported in world literature. Etiology remains unknown but has also been associated with bone marrow transplant patients, infection and autoimmunity. The median age of presentation is 53 years. Patient presents with a long history of persistent shortness of breath on exertion, dry cough. Around 1/3 patient presents with Pneumothorax. Patient is usually thin built. PFT reveals restrictive pattern. CT scan shows Upper and middle lung zone pleural and subpleural reticular parenchymal fibrosis with demarcation between affected and unaffected zone. Diagnosis is made clinically, radiologically and histologically.

KEYWORDS: Pleuro-parenchymal fibroelastosis (PPFE), Idiopathic interstitial pneumonia, Interstitial lung disease (ILD).

CASE REPORT
A 45-year-old female, nonsmoker and housewife presented to us in OPD with chief complains of shortness of breath for 3 years which was of MMRC-III grade, gradual onset, persistent and progressive in nature, with no diurnal, seasonal and postural variation. She also had dry and persistent cough for 3 years. There was h/o weight loss and loss of appetite. She denied any complains of hemoptysis, orthopnea, PND. H/o hypothyroidism for 5 years was under medication. No history of HTN, DM. No h/o surgery. No h/o ATT intake. Patient was a non-smoker. On general examination patient was thin built, pallor was present. On respiratory system examination, B/L inspiratory fine crept was present. Other systemic examinations were within normal limits.

INVESTIGATIONS
Complete blood count: HB- 11.5
TLC- 6510
DLC- N91L5
PLT- 2.12 lakhs
TSH-15
RFT, LFT, RBS, HBA1c, ECG, 2D-ECHO were within normal limit.

Table 1: 6 minute-walk-test

	SpO_2	Pulse rate	Distance	BORG scale
Before	90%	86 bpm		0
After	84%	120 bpm	300 meters	7

Digital CXR PA view

Figure 1: (A) lean and thin patient, (B) X-ray chest AP view showing bilateral diffuse reticulations along with areas of focal consolidation.

CECT thorax

- CT reveals fibrocystic and fibro-parenchymal changes along with reticulations and consolidations in bilateral lung fields with subpleural distribution, predominantly bilateral upper lobes.
- Small areas of honeycombing in left apical region, involving anterior segment of left upper lobe. Tractional bronchiectasis noted in bilateral lungs.
- All the features are suggestive of pleuro-parenchymal fibroelastosis.

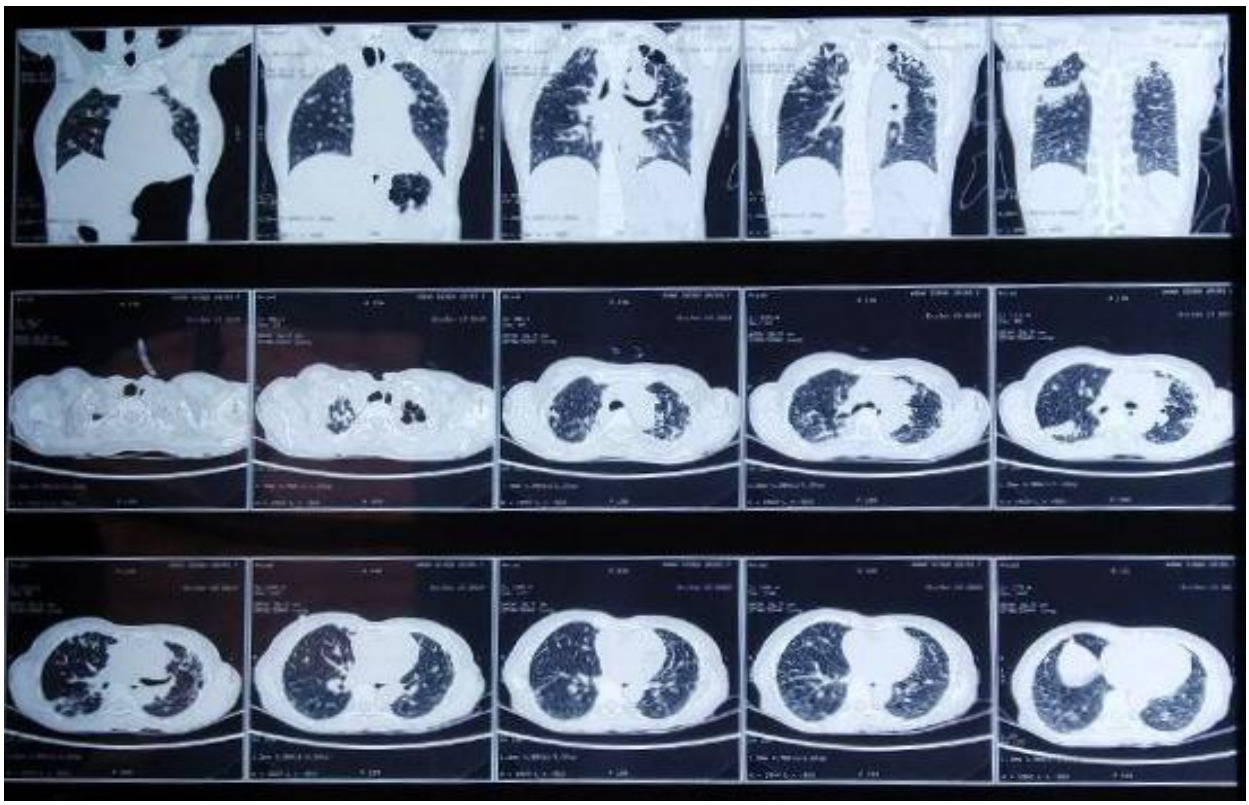

Figure 2: CECT-Thorax suggestive of pleuro-parenchymal fibroelastosis.

REFERENCES

1. Fishman's Pulmonary Diseases and Disorders, 5th ed. Editor-in-Chief: Michael A. Grippi. Editors: Jack A. Elias, Jay A. Fishman, Robert M. Kotloff, Allan I. Pack, Robert M. Senior. Video Editor: Mark D. Siegel. 278 contributing authors from 12 countries. New York: McGraw-Hill Medical; 2015. ISBN-13: 978-0071807289. ISBN-10: 0071807284
2. Frankel SK, Cool CD, Lynch DA, et al. Idiopathic pleuroparenchymal fibroelastosis: description of a novel clinicopathologic entity. Chest, 2004; 126: 2007–2013.
3. Becker CD, Gil J, Padilla ML Idiopathic pleuroparenchymal fibroelastosis: an unrecognized or misdiagnosed entity? Mod Pathol, 2008; 21: 784–787.
4. Keith C. Meyer, Steven D. Nathan, Editors, Idiopathic Pulmonary Fibrosis, A Comprehensive Clinical Guide, Second Edition 2019.