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CASE REPORT

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A middle-aged man with dyspnea and hoarseness: an unusual case of vocal cord paralysis

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Abstract

Background Pleuroparenchymal fibroelastosis (PPFE) is a rare and distinct form of Interstitial lung disease predominantly affecting the upper lung zones. It is characterized by fibrotic thickening of the visceral pleura and adjacent subpleural parenchyma. While common complications include spontaneous pneumothorax and pneumomediastinum, vocal cord paralysis (VCP) or paresis has been increasingly recognized as a potential manifestation in recent reports.

Case presentation We present a 49-year-old man presenting with progressive dyspnea, hoarseness, and left-sided vocal cord paralysis. Imaging studies revealed upper lobe-dominant fibrotic changes associated with significant pleural thickening consistent with PPFE. A comprehensive evaluation ruled out secondary causes of PPFE and other potential etiologies of VCP. Despite supportive management, the patient's VCP persisted, likely due to architectural distortion of the lung affecting the recurrent laryngeal nerve pathway.

Conclusions This case adds to the limited but growing body of literature on the association between PPFE and VCP. Understanding this rare complication is crucial for early recognition and appropriate management, as it highlights the diverse clinical manifestations of PPFE and its impact on patient outcomes.

Keywords Pleuroparenchymal fibroelastosis, Vocal cord paralysis, Interstitial lung disease, Recurrent laryngeal nerve, Tracheobronchial distortion

Background

Pleuroparenchymal (PPFE) is a rare form of interstitial pneumonia characterized by an upper lung zone-predominant fibrotic process involving the visceral pleura and adjacent subpleural parenchyma [1]. While the disease is largely idiopathic, non-idiopathic cases of PPFE have increasingly been reported in association with other interstitial lung diseases, including idiopathic pulmonary fibrosis, hypersensitivity pneumonitis (HP), and familial pulmonary fibrosis [1]. In clinical practice, although the definitive diagnosis relies on pathological evidence, this may not always be feasible due to the disease's rapid progression and the risk of respiratory complications

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following bronchoscopic or surgical lung biopsy. Therefore, radiological criteria are often used for diagnosis [2]. Similar to other idiopathic interstitial pneumonias (IIPs), dyspnea on exertion and dry cough are the primary symptoms [1–3]. While spontaneous pneumothorax and pneumomediastinum are the most common complications, recent reports have also described cases of VCP or paresis [3]. This report describes an unusual case of IPPFE presenting with left sided VCP.

Case presentation

A 49-year-old man was referred to our clinic by an otolaryngologist due to abnormal lung HRCT scan. He had been experiencing progressive shortness of breath for the past year. Initial evaluation by a local pulmonologist included a CT scan and spirometry, which revealed fibrotic changes in the lungs and a restrictive pattern on spirometry. However, due to inadequate follow-up, a definitive diagnosis was not established.

In the preceding two months, the patient developed new-onset hoarseness in addition to worsening shortness of breath. He consulted an otolaryngologist, and video laryngeal stroboscopy (VLS) demonstrated left-sided VCP. To investigate the underlying cause of vocal cord paralysis, further imaging studies, including a neck and chest CT scan and brain MRI, were performed. The brain MRI and neck CT were unremarkable, but the chest CT revealed bilateral upper lobes pleural thickening, parenchymal fibrosis, traction bronchiectasis, tracheal dilation and rightward deviation (Fig. 1a-b). The patient was subsequently referred to our center for further evaluation.

The patient was tall (182 cm) and underweight (55 kg). Physical examination revealed suprasternal notch deepening and platythorax (flattening of the chest cage with reduced anteroposterior diameter). His oxygen saturation was 92% while breathing room air. He had a history of smoking and opium use, both discontinued three years prior. There was no significant past medical history, occupational exposure, or drug history.

Fiberoptic bronchoscopy confirmed left-sided vocal cord paralysis (Fig. 2) and rightward tracheal deviation, without mucosal abnormalities. Bronchoalveolar lavage cell differential was normal, and testing for *Mycobacterium tuberculosis* and *Aspergillus* via PCR was negative.

There was no clinical or serological evidence of connective tissue disease or vasculitis. Based on typical imaging findings and the exclusion of competing diagnoses, VCP secondary to primary PPFE was considered. Body plethysmography showed a restrictive pattern with an elevated residual volume (RV)/total lung capacity (TLC) ratio, a characteristic feature of PPFE (Table 1).

Six months after the initial visit, the patient continued to complain of hoarseness and progressive dyspnea on exertion.

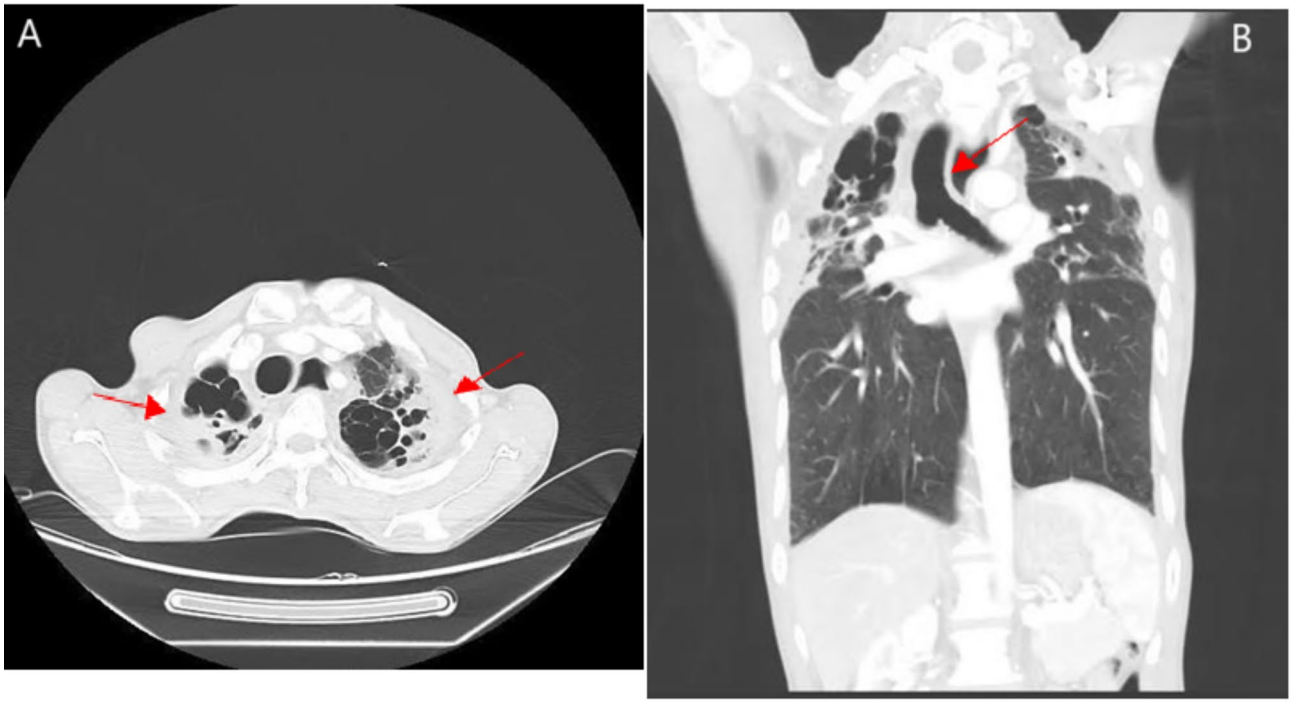

Fig. 1 **A**. Chest CT scan (axial view). Bilateral upper lobe pleural thickening (red arrow), parenchymal fibrosis and traction bronchiectasis. **B**. Chest CT scan (coronal view). Tracheal dilation and rightward deviation (red arrow) due to architectural distortion of both upper lobes

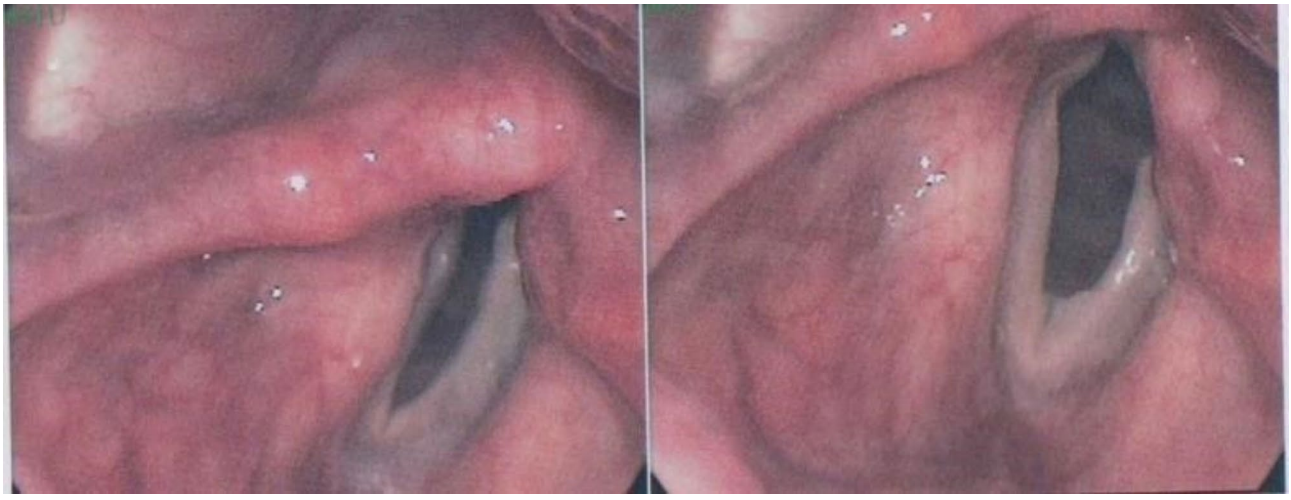

Fig. 2 Left vocal cord paralysis without mucosal abnormalities on fiberoptic bronchoscopy

Table 1 Summary of patient's body plethysmography and DLCO. FEV1(liter): forced expiratory volume in one second, FVC(liter): forced vital capacity, TLC(liter): total lung capacity, RV(liter): residual volume, DLCO(mmol/kpa/minute): lung diffusion capacity for carbon monoxide

Parameter	Predicted	Actual	Percent %
FEV1	4.00	1.51	38
FVC	5.16	1.70	33
FEV1/FVC		0.84	
TLC	7.46	4.89	66
RV	2.17	3.10	143
RV/TLC	32	63	199
DLCO	11.16	5.86	52

Discussion

The primary etiologies of VCP involve conditions that affect the recurrent laryngeal nerve (RLN), including tumors of the thyroid, esophagus, mediastinum, and pleura; tuberculosis; iatrogenic causes after cervical or mediastinal surgery; diabetes mellitus; mitral valve stenosis; and rapid swelling of the pulmonary artery. In rare instances, some paranchymal lung disease such as coal workers' pneumoconiosis with massive fibrosis, lung collapse due to cystic fibrosis, and invasive pulmonary aspergillosis, idiopathic or secondary mediastinal fibrosis may also result in VCP [4–9]. Typically, the left RLN is more often affected, as its path is longer, as observed in this case [10, 11].

Although the mechanism of RLN paralysis in lung diseases remains unclear, recent reports propose several mechanisms. These include stretching or retraction of the RLN due to chest wall adhesion resulting from pleural fibrosis [7], distortion of the tracheobronchial tree leading to RLN traction or compression [3, 11], and left-sided RLN compression at the aortopulmonary window, where the aortic arch and left pulmonary artery are in close proximity [3, 7, 11]. Lardinois et al. reported a case of left RLN paralysis associated with silicosis, where progressive recovery of the voice was observed 15 weeks after careful dissection of the nerve and release from scar encasement during video mediastinoscopy [6].

To date, seven cases of PPFE with VCP have been reported, including the present case. In all but one case, the paralysis involved the left vocal cord [11]. Dominant left RNP along right tracheal deviation, support RLN traction or compression as most probable mechanism of vocal cord paralysis in PPFE. After excluding treatable and more dangerous cause of new onset hoarseness, VCP secondary to parenchymal involvement should be considered in PPFE patient.

Clinicians encountering patients with upper lobe-predominant fibrotic lung disease and concurrent VCP should include PPFE as a differential diagnosis.

Abbreviations

PPFE	Pleuroparenchymal fibroelastosis
VCP	Vocal cord paralysis
IPPFE	Idiopatic pleuroparenchymal fibroelastosis
RLN	Recurrent laryngeal nerve
HP	Hypersensitivity pneumonitis
IIPs	Idiopathic interstitial pneumonias
CT	Computed tomography
MRI	Magnetic resonance imaging
VLS	Video laryngeal stroboscopy
PCR	Polymerase chain reaction
RV	Residual volume
TLC	Total lung capacity
BAL	Bronchoalveolar lavage

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Author contributions

SNT analyzed and interpreted the patient's clinical data and contributed to drafting the manuscript. MA assisted with patient management, performed the literature review, and contributed to manuscript preparation. MJF

supervised the clinical aspects of the case, provided critical revisions, and served as the corresponding author. All authors read and approved the final manuscript.

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Data availability
The raw data analyzed in this study are available from the corresponding author, Dr. Mohammad Javad Fallahi (Email: falahijavad@yahoo.com), upon reasonable request.

Declarations

Ethics approval and consent to participate
Written informed consent was obtained from the patient for participation and publication of this case report.

Consent for publication
Written informed consent was obtained from the patient for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare no competing interests.

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