Case Report

A case report on pulmonary fibro-emphysema: an incorrectly framed case

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Abstract: This case report describes the combination of pulmonary fibrosis and emphysema (CPFE) as a possible new addition to the growing list of smoking-related lung diseases, which are characterised by the coexistence of usual interstitial pneumonia (UIP) or non-specific interstitial pneumonia (NSIP) with emphysema in tobacco smokers.

Keywords: COPD, Therapy, Spirometry.

Introduction

Combined pulmonary fibrosis and emphysema (CPFE) is a new addition to a growing list of smoking-related lung diseases, characterised by the coexistence of idiopathic pulmonary fibrosis (IPF) with emphysema in tobacco smokers[1]. CPFE accounts for between 5 and 10% of cases of diffuse interstitial lung disease[2].

Three distinct CPFE patterns have been described in the recent literature:

- Progressive transition with diffuse emphysema (centrilobular and bullous) and bubble/honeycomb transition zone
- Paraseptal emphysema with a predominance of subpleural bubbles of increasing size at the bases
- Separate processes with independent areas of fibrosis and emphysema.[3]

Clinical Case

A 77-year-old patient, came to my attention for reported episodes of dry cough, dyspnoea associated with desaturation under minimal exertion, and productive cough with a light-yellowish plug. The patient also reported a persistent "air hunger" sensation.

Remote pathological history: former building contractor, occupational exposure to cement dust, adverse reaction to theophylline with diarrhoea, former heavy smoker (90Pack/year), Body Mass Index: 25. Emphysematous chronic obstructive pulmonary disease (COPD) with hypoxaemic respiratory failure in long term oxygen therapy (OTLT) and inhaled corticosteroid/long-acting β2-agonist (ICS/LABA). Chronic heart failure in previous ischaemic heart disease, previous lung cancer undergoing left lobectomy in 2015, diabetes mellitus type II, systemic arterial hypertension. Up to the time of the visit, he had been treated by a pulmonologist colleague for COPD with aerosol therapy, ICS/LABA, and oral corticosteroid treatment (OCS). He performed spirometry with lung diffusing capacity test (DLCO) and computer tomography (CT) scan, whose results are reported in Figure 1.

Figure 1. Spirometry of the patient

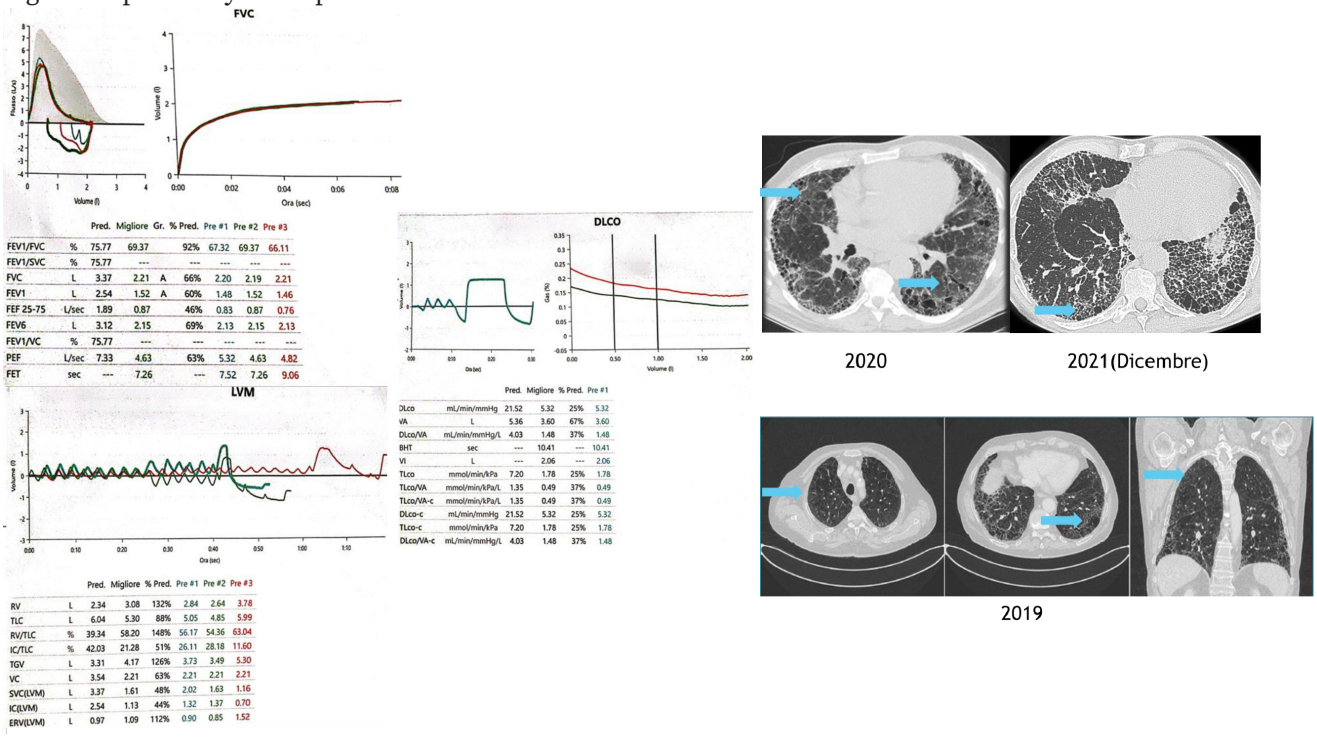

Figure 1 shows on the right the presence on global spirometry with DLCO the presence of moderate degree obstructive deficit with severe reduction of DLCO and presence of high ratio of RV to TLC (148% of predicted) and RV: 132% low tidal volume and inspiratory capacity (VT: 63% of predicted, IC(LVM): 44% predicted). The CT scan of the chest shows a progressive course of the disease resulting in the presence of distortion of the lung parenchymal architecture with prevalence at the apices of paraseptal emphysema bubbles and at the bases the presence of honeycombing with subpleural sparing, traction bronchiectasis and areas of bilateral GGO that at the left base converge into thickening (December 2021). Therefore, the picture appeared to be suggestive of CPFE at first hypothesis.

Legend*: DLCO = diffusing capacity for carbon monoxide; CT = computed tomography; RV = residual volume; TLC = total lung capacity; VT = vital capacity; IC = inspiratory capacity; LVM = Lung Volume Measurements; GGO = Ground glass opacity; CPFE = Combined Pulmonary Fibrosis and Emphysema.*

Aim

The aim of this case report is to highlight the correct approach to the diagnosis of complex pathologies, which in most cases are discussed with a multidisciplinary team. However, the expertise of a pulmonologist can change the diagnostic-therapeutic approach to complex pulmonary pathology, and this has been gaining in value in recent years, also in relation to the increased difficulty and complexity of certain pathologies such as this, which are not very common in the general population. Furthermore, it is important to value the step-by-step approach in the diagnosis of these pathologies, integrating various instrumental examinations that our structure has available.

Diagnostic-Therapeutic Approach

The diagnostic and therapeutic approach used is based on the ATS 2018 document on the diagnosis of fibrotic disorders, however, as this initial hypothesis was made at a private clinic, the patient was subsequently referred to a regional reference centre that confirmed the diagnostic hypothesis.

Anti-fibrotic therapy was started immediately as the fibrotic pathology was progressive over time with distortion of the parenchymal architecture and with damage leading to significant symptoms reported by the patient. In the first instance, the patient had been labelled as COPD and the therapeutic approach was incorrect as DLCO and global spirometry had not been performed. These examinations allow the specialist to have fundamental information to set eventual therapy and need to be more developed in our area within public structures.

Discussion

This case report highlights the importance of correctly diagnosing the idiopatic pulmonary fibrosis in the context of a COPD, which has an incidence of 5-10% within fibrotic pathologies. This is important because it shows how the symptoms and clinic of COPD patients can sometimes also underlie a fibrotic pathology. This case report also emphasises the importance of an early global spirometric examination with DLCO in patients with a high degree of dyspnoea, since it can correctly classify and diagnose the COPD. Notably, CODP and idiopatic pulmonary fibrosis can sometimes be confused given that they have common symptoms, including minimal exertional dyspnoea; however, high-resolution CT scans, together with spirometry remain the cornerstones for the diagnosis of these pathologies; lastly, use of Pirfenidone as an early antifibrotic therapy is also emphasised to slow down the progression of the fibrosis.

Conclusions

The patient's clinical and radiological features supported a CPFE diagnosis, with radiological features of emphysema, which was predominant in the upper zone, and pulmonary fibrosis of the lower lobes, which radiologically, had the characteristics of a UIP.

In this case, the radiological features described a UIP pattern, and a diagnosis of idiopathic pulmonary fibrosis was made, so the patient started Pirfenidone therapy, triple inhaled treatment with ICS/LABA plus a long-acting muscarinic antagonist (LAMA), and follow-up with CT scan once a year.

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