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Clinical Case Seminar **A4(1-5)**

An unusual cause of dyspnoea in a patient with chronic ischemic heart disease

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Abstract

A 70-year old male was referred to our attention for worsening dyspnea in the last three months. His past medical history was characterized by an acute myocardial infarction and an episode of ventricular tachycardia with positioning of an implantable cardioverter defibrillator. At the physical examination “velcro” inspiratory crackles were present, however lung volumes, diffusing capacity of the carbon monoxide and arterial blood gases values were within the normal limits, whereas the computed tomography of the chest showed a typical pattern of idiopathic pulmonary fibrosis. Long-term treatment with pirfenidone was started.

Conclusions: Idiopathic pulmonary fibrosis is a rare chronic, progressive lung disease that may present with progressive dyspnea which may be confused with cardiogenic dyspnea.

KeyWords:: Idiopathic Pulmonary Fibrosis; dyspnea; heart failure.

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Introduction

Idiopathic pulmonary fibrosis (IPF) is a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause occurring in adults (1).It is usually chronic and progressive, with characteristic imaging and histologic appearances, that occurs primarily in older adults and is often fatal. This disease is frequently misdiagnosed by internal medicine specialists and primary care physicians (2). Cardiac comorbidities are often associated with IPF and the differential diagnosis may be difficult (3).

Case report

The case concerns a 70-year-old, formerly employed as a bakery, who presented to our attention

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for worsening dyspnea in the last three months. He was a former smoker (he quitted in 1998) of 25 pack-years, with a long history of systemic arterial hypertension [treated with ramipril (10 mg daily)] and diabetes mellitus [treated with metformin (400 mg twice daily) and glibenclamid (2.5 mg twice daily)] and dyslipidemia [treated with rosuvastatin (10 mg daily)]. He had an anxiety depressive disorder treated with lorazepam (1 mg daily).His past medical history was characterized by an acute myocardial infarction (30 years before the presentation) and (2011) an episode ventricular tachycardia with positioning of an implantable cardioverter defibrillator.At the hospital admission, the systemic blood pressure was 135/75 mmHg with a pulse frequency of 96 bpm (rhythmic) and a body axillary temperature of 36°C. His height was 163 cm with a weight of 84 Kg and a body mass index of 31 Kg/m^2. Physical examination of the chest revealed the presence of bilateral "velcro" inspiratory crackles on basal lung fields at lung auscultation. No other physical signs outside the chest were pathological. Routine laboratory tests were normal. A panel of serum auto antibodies is presented in **Table 1.**

Table 1. Panel of serum auto antibodies

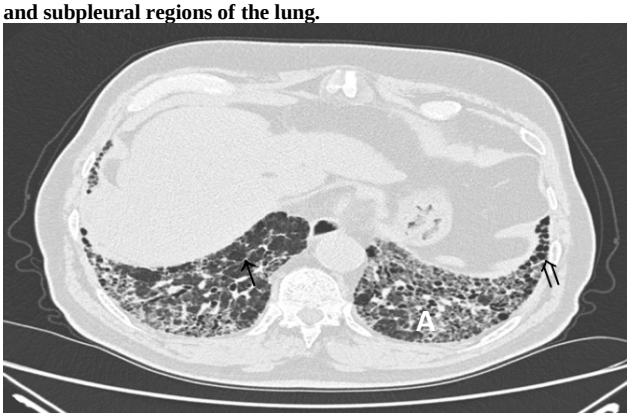
Parameters	Value	Normal Range
ANA	1:160	≤ 1:80
ASMA	1:80	negative
Rheumatoid factor	7 IU/ml	0-14 IU/ml

ANA:antinuclear antibodies; ASMA:anti-smooth muscle antibodies

Lung volumes, diffusing capacity of the carbon monoxide and arterial blood gases values were within the normal limits **(Fig.1).**

The six-minute walk test showed a distance of 175 meters (23% of the predicted value) associated with oxygen desaturation (minimum value of SpO_2=90%) and dyspnea (score 2 of the visual Borg scale 1-10 where ten was the most intense degree). The echocardiography revealed a reduced ejection fraction (50%; nv > 55%) and normal pulmonary artery systolic pressure. The high resolution computed tomography of the chest showed a typical pattern of idiopathic pulmonary fibrosis according to the Fleischner Society criteria (Fig.2) (4). Further more invasive diagnostic tests were not performed.

Fig. 2.High resolution computed tomography of the chest showed a typical UIP characterized by the presence of honeycombing (open arrow) and reticular pattern (closed arrow) located in the basal and subpleural regions of the lung.



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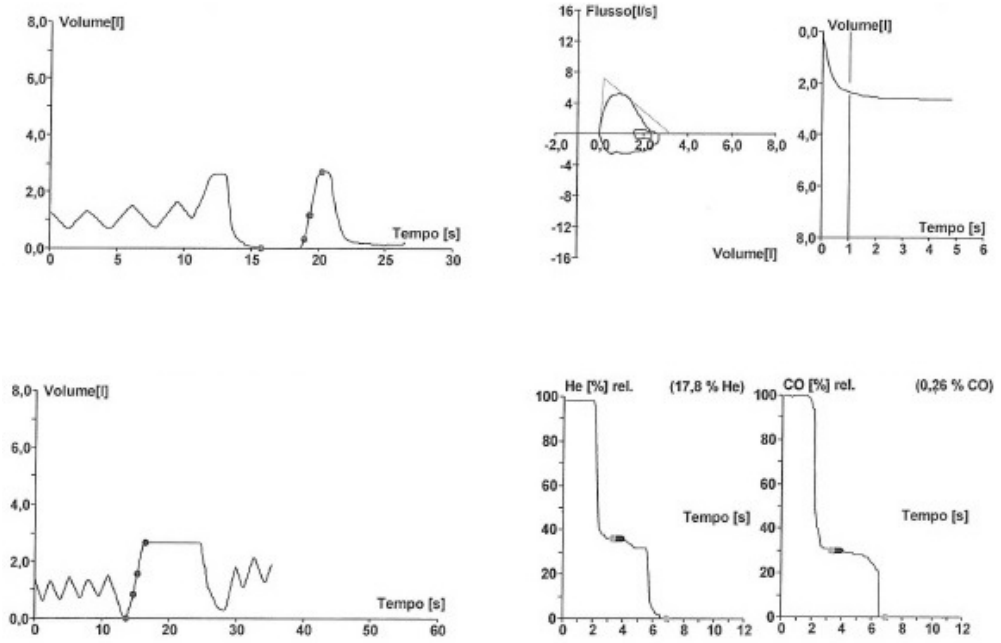
Long-term treatment with pirfenidone (267 mg three times daily) and omeprazole (40 mg daily) was started together with long-term follow-up of the patient according to current guidelines (1).

Discussion

IPF is the most common type of idiopathic interstitial pneumonia (IIP), that is more common in male smokers ≥50 years old (5). The most favored conceptual model of the pathogenesis of IPF posits that recurrent, subclinical epithelial injury superimposed on accelerated epithelial aging leads to aberrant repair of the injured alveolus and deposition of interstitial fibrosis by myofibroblasts (2). Every clinician should consider this interstitial lung disease in the differential diagnosis of all adults presenting with unexplained exertional dyspnea typically progressing over a period of month to years and associated with chronic dry cough and/or velcro-like crackles on

Figure 1.Lung volumes, diffusing capacity for the carbon monoxide and arterial blood gases values were within the normal limits



Parameters	Unit	Ref	Pre	Pre%Ref
FEV1	Liters	2.49	2.32	93
FVC	Liters	3.22	2.60	81
FEV1/FVC	%	75	85	
FEF25-75%	L/sec	2.86	3.69	129
PEF	L/sec	7.16	5.17	72
TLC-He	Liters	5.95	4.79	80
FRC-He	Liters	3.36	2.80	83
RV-He	Liters	2.44	1.94	79
RV/TLC-He	%		42	
TLCO	mmol/min/kPa	7.47	6.20	83
KCO	mmol/min/kPa/L	1.30	1.40	108
pH		7.35-7.45	7.44	
PaO_2	mmHg	80-100	85	
$PaCO_2$	mmHg	35-45	35.8	
Serum bicarbonates	mmol/L	22-26	24	
SaO_2	%	94-100	96	

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examination (2). In fact, the lung auscultation of basal bilateral velcro-like crackles has an excellent sensitivity and good specificity for the diagnosis of IPF (6). These sounds are different from adventitious sounds associated with heart failure (7).A detailed medical history is of fundamental importance for the differential diagnosis with other chronic secondary fibrosing lung disease, such as chronic hypersensitivity pneumonitis and pneumoconiosis (4). When the computed tomography of the chest pattern is indicative of typical or probable UIP (usual interstitial pneumonia) the diagnosis is confident (4). Whereas in the presence of a CT pattern indeterminate for UIP, more invasive diagnostic tests (including lung biopsy) may be considered, according the general clinical conditions of the patient and his/her preferences (4). The incidence of exacerbations in the patients with IPF is variable with a 1 and 3-years incidence raging between 8.6% and 23.9% (8). Due to the current absence of effective therapies and the high mortality (median life expectancy of 2.2 months) a new definition of IPF exacerbation has been proposed including all causes of respiratory deterioration except for acute heart failure and volume overload (9). The majority of patients with IPF have comorbidities, particularly gastro-oesophageal reflux (prevalence ranged from 5% to 94%), that may result in more frequent exacerbations, rapid decline of lung function and reduced survival (3).Currently in Italy there are two drugs (nintedanib and pirfenidone) approved for the long-term treatment of IPF both have shown to significantly reduce the decline of lung function (forced vital capacity) and the number and severity of exacerbations leading to hospitalization, but do not reduce significantly the mortality (2). Lung transplantation can improve survival among patients with IPF so transplant discussion was recommended at the time of diagnosis (10). In conclusion, idiopathic pulmonary fibrosis is a rare chronic, progressive lung disease that may present with progressive dyspnea which may be confused with cardiogenic dyspnea.

Conflicts of Interest: There is no potential conflict of interest, and the authors have nothing to disclose. This work was not supported by any grant.

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