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Challenges in IPF diagnosis, current management and future perspectives: Patient case 2

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Patient presentation and diagnosis

The patient is a 73-year-old male retired house painter and ex-smoker who was admitted to the respiratory medicine department of a district hospital in Greece due to the presence of streaks of blood in his sputum. His past medical history included coronary artery disease, arterial hypertension and supraventricular tachycardia (SVT). A high resolution computed tomography (HRCT) was performed which showed enlarged mediastinal lymph nodes with evidence of fibrosis (Figure 1).

The initial diagnosis was acute bronchitis with underlying fibrosis (with no further information about the nature of the fibrosis). The diagnostic work-up had taken place in a district hospital where both radiologists and respiratory physicians had very limited knowledge of interstitial lung diseases. The patient was treated with antibiotics and low dose of oral prednisolone (10 mg) for 2 months.

Six months later he was admitted to the cardiology department of a different hospital because of SVT and a respiratory consultation was undertaken due to his past medical history of fibrosis. Careful examination revealed progressive shortness of breath over the past 10 months, mild dry cough and chest auscultation revealed inspiratory crackles in the mid-lower lung fields. HRCT images have been reviewed and were suggestive of a possible usual interstitial pneumonia (UIP) pattern. On multidisciplinary discussion the combination of clinical and CT findings together with the patient's age suggested a diagnosis of idiopathic pulmonary fibrosis (IPF) (1).

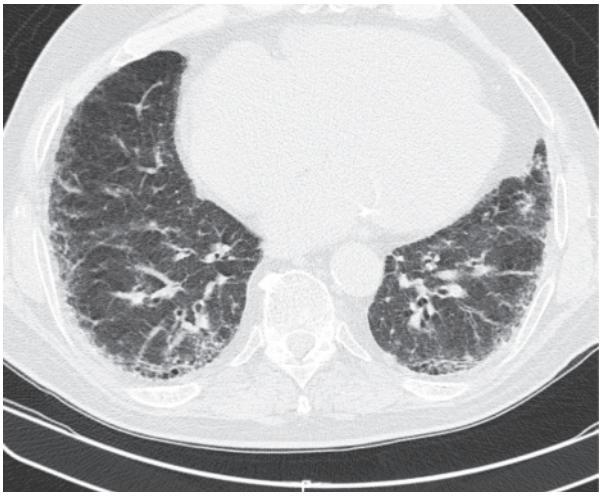

Fig. 1. Presenting HRCT

Management and follow-up

The patient was started on pirfenidone. After taking the full dose (2403 mg per day) for 7 months his condition stabilised and he resumed gardening and walking for exercise. Although he had been educated on strategies to prevent the development of pirfenidone-associated side effects, particularly since he lived in a sunny climate, the patient nonetheless developed a photosensitivity reaction on his head, ears and both hands. The skin-related reaction resolved following pirfenidone discontinuation and treatment

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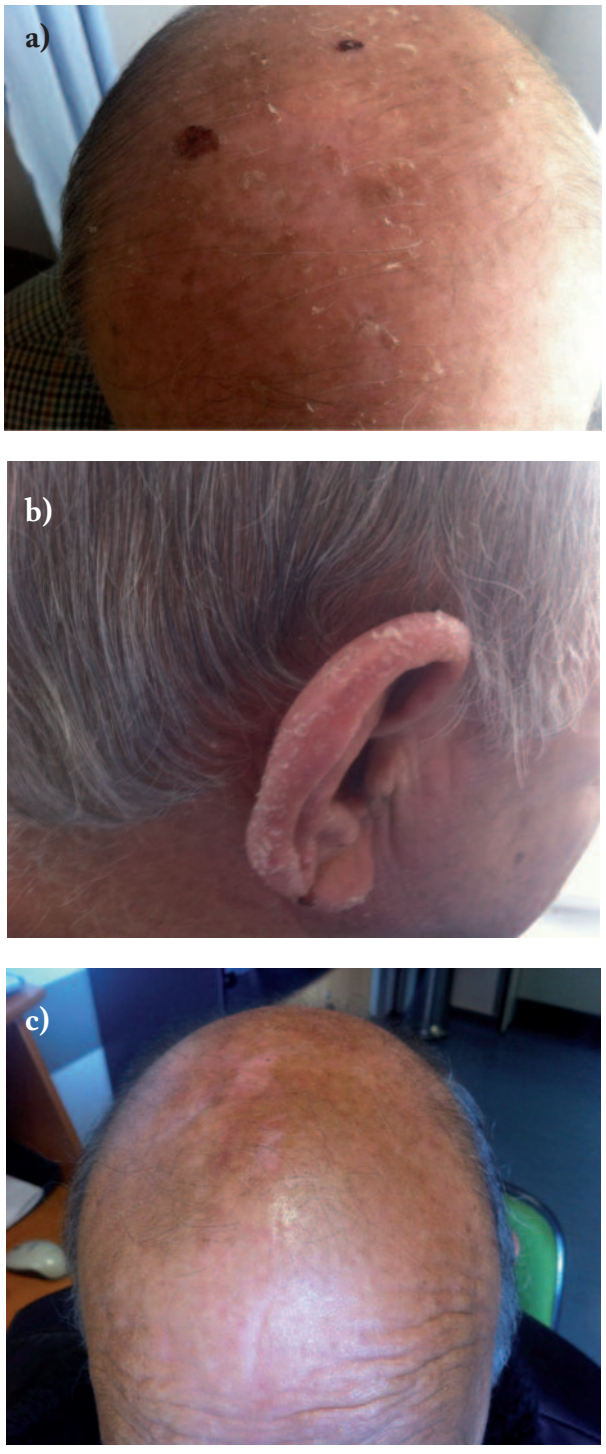

Fig. 2. a, b) Photosensitivity reaction on areas of sun exposure. c) Resolution of skin reaction after discontinuation of pirfenidone for two weeks and very slow re-titration starting with 1 tablet per day and increase of the dose by 1 tablet per week

for 15 days with low dose (15 mg) of oral steroids. Subsequently treatment has been restarted at a reduced dose (two tablets 3 times daily). Despite taking protective measures, he presented again with rash on sun-exposed body areas. Pirfenidone was again discontinued until the skin reaction resolved and then re-introduced after 3 weeks but very slowly (starting with one tablet per day and increasing the dose by one tablet every week) (Figure 2). He is currently taking two tablets twice a day and is clinically and functionally stable with no further skin reactions.

Conclusion

This case, in which diagnosis of IPF was delayed, illustrates the need to promote greater awareness of IPF in local hospitals. After the onset of exertional dyspnoea, delayed referral to a tertiary care centre is associated with increased mortality independently of disease severity. If findings are suggestive of interstitial lung diseases and IPF is in the differential diagnosis, patients should be immediately referred to a specialist centre (2).

The case also reflects the findings of the 3 major pirfenidone trials in which rash and photosensitivity were among the most common adverse events (3,4). This patient's reaction was reversible with temporary discontinuation, treatment with steroids and slow re-titration, but highlights the need for patients on pirfenidone to avoid exposure to the sun as much as possible along with taking appropriate preventative measures (5).

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