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Challenges in IPF diagnosis, current management and future perspectives: Patient case 1

Jim Egan
Department of Respiratory Medicine, Mater Misericordiea University Hospital, Dublin, Ireland

Patient presentation and diagnosis

A 78-year-old female was referred to our department in April 2011 with shortness of breath. Clinical examination revealed bilateral limited crackles but no signs of finger clubbing. Lung function evaluation showed a percent predicted FVC of 78 and DLco of 67. The patient underwent HRCT imaging which showed cardinal features of a UIP pattern enabling a rapid diagnosis of IPF without the need for surgical lung biopsy (Figure 1).

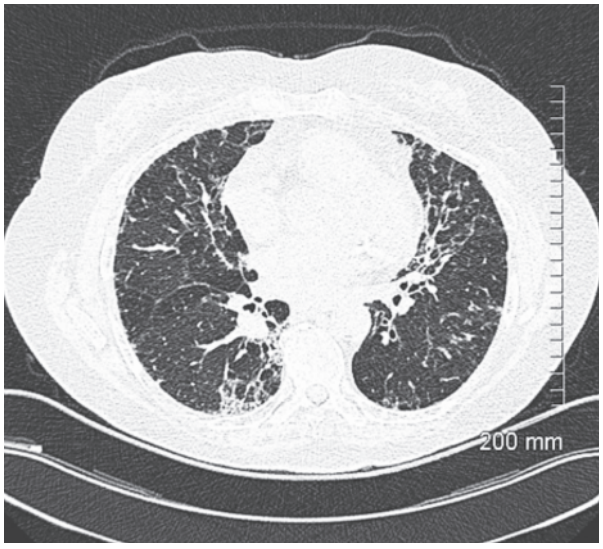

Fig. 1. Diagnostic UIP pattern on HRCT

Correspondence: Jim Egan
Department of Respiratory Medicine, Mater Misericordiea University Hospital, Dublin, Ireland

Management and follow-up

Until recently, there were no approved therapies for patients with IPF and, historically, prognosis in these cases was variable with a median overall survival of just 2–5 years (1,2). At the time of this patient's diagnosis pirfenidone had just recently been approved by the European Medicines Agency for the treatment of patients with mild-to-moderate disease but was not widely available in all European countries. However, it was available for compassionate use through a European pirfenidone named patient programme (NPP) in a number of countries to help patients who could most benefit from this new treatment. We enrolled this patient in the pirfenidone NPP and she was titrated over 2 weeks to receive pirfenidone 2403 mg/d. Follow-up over the subsequent 3 years has shown long-term stabilisation of lung function without significant decline enabling the patient to improve her mobility with a hip replacement operation in 2012 and, ultimately, outliving her husband (Table 1).

Table 1. Lung function parameters and events during three-year follow-up on pirfenidone treatment

Timeline	% predicted FVC	% predicted DLco
30 November 2011	FVC 80%	DLco 62%
29 February 2012	FVC 82%	DLco 69%
30 May 2012	FVC 81%	DLco 69%
29 August 2012	FVC 81%	DLco 62%
Hip replacement 28 November 2012	FVC 73%	DLco 63%
23 October 2013	FVC 79%	DLco 62%
12 February 2014	FVC 75%	DLco 55%
14 May 2014	FVC 77%	DLco 60 %

Conclusion

This patient case underscores the importance of early diagnosis of IPF and early treatment with pirfenidone to reduce decline in lung function and potentially improve the current prognosis of this disease. In this case, early treatment of IPF with pirfenidone has resulted in stabilization of lung function over a three-year period.

References

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