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PIRFENIDONE IN IDIOPATHIC PULMONARY FIBROSIS: A CASE REPORT

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BACKGROUND:

Idiopathic pulmonary fibrosis (IPF) is a chronic, limited lung fibrosing interstitial pneumonia of unknown cause with poor prognosis and few therapeutic options, with a median survival of 2-5 years from diagnosis. It is characterized by fibroblast proliferation and abnormal accumulation of extracellular matrix molecules, particularly collagen fibers. Pirfenidone is the first IPF-specific antifibrotic, anti-inflammatory and antifibrotic properties whose mechanism of action has not been fully established. To date, pirfenidone is the only drug with proven efficacy in the treatment of IPF. It is considered an orphan drug, and not yet marketed in all European countries, so that additional monitoring is required

OBJECTIVE:

To describe the evolution of the patient treated with pirfenidone as well as the safety of this new treatment.

METHODS and STUDY DESIGN:

A prospective observational study was conducted. The patient, a 65 year old male, is ex-smoker since 2 years ago (with an index of cumulative tobacco consumption of 57 packs / year). He was diagnosed with IPF in 2012 by clinical and radiological criteria. Pirfenidone was approved as a foreign medication for a period of three months by the Ministry of Health.

RESULTS:

After titration regimen, usual pirfenidone dose was administered (2403 mg/daily) After a treatment period of 3 months, forced vital capacity (FVC) experienced less than 10% decrease (only 1.2%) and diffusing capacity for lung carbon (DLCO) decreased 6.1% showing no radiological progression. There has been no increase in transaminases, neither digestive disturbances neither weight loss.

FUNCTIONAL ANALYSIS	
Before Pirfenidone (month -2)	FVC 2730 (73,7%) DLCO 40,6
After Pirfenidone (month +3)	FVC 2690 (72,8%) DLCO 38,1%

Months after pirfenidone	*LDH (240-480 U/L)*	*GOT (0-40 U/L)*	*GPT (0-41 U/L)*	*PA (40-129 U/L)*
+1	351	17	11	-
+2	376	16	9	52
+3	350	17	18	46
+4	323	19	16	-

CONCLUSIONS:

→ Pirfenidone has been used successfully to date in our case, so it is possible to continue the treatment until a new patient reevaluation at 6 months in order to prevent lung transplant.
→ Althought more number of treatments and longer ones are needed, pirfenidone seems to be a well-tolerated treatment option for the IPF

REFERENCES:

http://www.ema.europa.eu/docs/es_ES/document_library/EPAR__Product_Information/human/002154/WC500103049.pdf