Characterization of Patients with Interstitial Pneumonia with Autoimmune Features (IPAF) and Its Comparison with Patients with Scleroderma-Related Interstitial Lung Disease and with Idiopathic Fibrosis

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Background: Diffuse parenchymal pulmonary diseases, called interstitial lung diseases, are a heterogeneous group of disorders that are classified together due to clinical, radiographic, physiological or similar pathological manifestations. The diagnosis of idiopathic interstitial pneumonias requires the exclusion of known causes of interstitial pneumonia. Identifying an underlying etiology is important for clinical perspectives because it impacts prognosis and treatment. A recent number of studies has shown that many patients diagnosed as idiopathic interstitial pneumonia have clinical elements that suggest an underlying autoimmune process without meeting established diagnostic criteria for connective tissue disease.

Objectives: Our objectives were characterize the clinical findings of patients who meet the IPAF criteria and compare them with the clinical characteristics of patients with scleroderma-related interstitial lung disease and patients with idiopathic pulmonary fibrosis.

Methods: We retrospectively reviewed 254 patients hospitalized at the Hospital Clínico de La Universidad de Chile between January 2012 and June 2018 who had ICD-10 diagnosis of J.84 (Other respiratory diseases principally affecting the interstitium) and J99.1 (Respiratory disorders in other diffuse connective tissue disorders). The electronic medical record was reviewed retrospectively to extract pertinent data. We applied IPAF criteria to this 254 patients. We then characterized the clinical, serological and morphological features of the IPAF cohort and compared outcomes with other ILD cohorts: scleroderma-related interstitial lung disease and idiopathic pulmonary fibrosis (IPF).

Results: Of 254 patients screened, 17 patients met the IPAF criteria. Mean age was 60 years with a female predominance. The most frequent pattern by high-resolution computed tomography was NSIP present in 46.7%. The median of Forced Vital Capacity was 82%, and median of DLCO was 50%. 14 patients (82%) were treated with corticosteroids. 11 patients (64%) used other immunosuppressant: 6 patients azathioprine, 4 mycophenolate and 1 patient used cyclophosphamide. One patient received a lung transplant in IPAF cohort. We identified 2 deaths in IPAF cohort, 6 in sclerosis systemic and 30 in IPF cohort. IPAF cohort survival was worse than Scleroderma cohort and better than the IPF cohort.

Conclusion: Our IPAF cohort is similar to the cohorts described in other studies, in relation to the age of diagnostic, female predominance and High-Resolution Computed Tomograph pattern. Also the trend in survival was similar to others previously described. Our study have limitations, the first one is related to the retrospective nature of the reviewed cohorts. Further prospective studies should be conducted for a more comprehensive evaluation of the evolution of these diseases and the impact of the treatments used.

REFERENCES

Disclosure of Interests: Karen Vergara: None declared, Silvana Saavedra: None declared, Felipe Reyes: None declared, Annelise Goecke Consultant for: Roche, Abbvie, Novartis, Pfizer, Paid instructor for: Roche, Speakers bureau: Roche, Novartis, Abbvie, Pfizer, Caterina Chesta: None declared, Sebastian Chavez: None declared