Background: Dermatomyositis (DM) is an idiopathic inflammatory myopathy. The recent years has increased its knowledge thanks to best characterization of myositis-specific antibodies that correlate with different clinical phenotypes with different clinical phenotypes.

Objectives: To describe the clinical and analytical features of a series of dermatomyositis: clinical debut, clinical manifestations as well as the treatments received and the evolution of the disease.

Methods: Patients diagnosed of dermatomyositis in a tertiary hospital between the years 1978-2018 according to the criteria of Bohan and Peter (1975) and according to Dalakas' classification criteria (2015). Clinical, analytical and immunological profile data were collected, as well as treatments received and the evolution of the disease.

Results: A total of 59 inflammatory myopathies diagnosed between the years 1985 and 2018 were included. 46 were dermatomyositis (78%), 9 polymyositis (15%) and 4 necrotizing myositis (7%). 69% were women and 31% were smokers. Clinic started at 54 ± 17 years. The initial manifestation was pulmonary in 28.7% followed by cutaneous manifestations at the same time. 45.5% behaved like a myopathic manifestation while 28% presented interstitial involvement. The most common non-specific interstitial pneumonitis (76%) followed by usual interstitial pneumonitis (17%). In these patients, DLCO was decreased (mean of 58.9% and 13.2%) as well as the FVC (average of 59%, 2.5L). 73.3% presented cutaneous involvement being the most common manifestations the Gottron papules (37.8%) and the heliotrope rash (35.6%), and up to 27% had cuticular affection. 64% had muscle involvement, affecting proximal and symmetrical. Neck flexors were affected in a 38% of patients while 20% had dysphagia. Only 3 patients presented dysphonia (7%) and 2 myocarditis.

Conclusion: 80% received corticotherapy at a dose of mg/kg/day and 20% required high doses of metildiprednisolone due to muscular involvement or pulmonary. 18% immunoglobulins and 11% cyclophosphamide. As maintenance 80% received disease modifying antirheumatic drugs (azathioprine 22%, methotrexate 15%, tacrolimus 13.9%, rituximab 11%) due to muscle (29.9%), cutaneous (24.4%) and pulmonary involvement (22.2%). As complications, 2 cases of the syndrome were registered hemophagocytic. Virtually all patients they presented pulmonary hypertension (mean 34 ± 12 mmHg). 5 patients (11.1%) were diagnosed with neoplasia, two of them after the diagnosis of DM. The mortality was 24%. 3 patients died due to a rapidly progressive pneumonitis, another 2 due to alveolar hemorrhage and three of them due to complication of the neoplastic disease. Dermatomyositis occurs in a variable way, with predominance of pulmonary, skin and muscle manifestations. They require corticotherapy and immunosuppressive treatment for maintenance, even so, mortality is high.

Disclosure of Interests: None declared