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General Properties and Prognosis of 22 NSIP Cases

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Objectives: Nonspesific interstitial pneumonia (NSIP) is an idiopathic interstitial pulmonary disease mostly effcting women between ages 40-50. Although known as idiopathic, NSIP is generally seen among patients with connective tissue disorders like systemic sclerosis, autoimmune myositis and hypersensitivity pneumonitis. NSIP has fibrotic, cellular and mixed types. Symptoms are nonspesific, it manifests with dyspnea and coughing. The most common radiographic findings on computer tomography (CT) are bilateral ground glass opacities and fibrotic infiltrates. The main treatment is corticosteroids. Our aim is to present general properties, diagnostic methods and prognosis of NSIP patients followed in our clinic.

Methods: All patients diagnosed as NSIP by either biopsy or radiology were enrolled. Patients' demographics, pulmonary function tests, treatments and prognosis were evaluated.

Results: Of the 22 patients, 14 were female. Mean age was 58,2 13 (14-81) and average duration of symptoms was 22.9 (5-98) months. 14(%62) of the patients had smoking history of average 18,7 packs/year. The most common symptoms were dyspnea and cough seen in 18 and 16 patients, respectively. None of the patients had a rheumotological disease except for 2 patients with Romatoid artritis and 1 with systemic sclerosis. 5 patients had history ov avian exposure. Mean pulmonary function tests were FVC: 1.96-0.79 (71%, 8225.2) FEV1: 1.87-0.74 (74%, 1825.9) FEV1/FVC: 85.5-8.9 DLCO: 49.95-19.38. Average 6-minute walking test was 33493,7 meters. Nine of the patients were diagnosed by transbronchial cryobiopsy; whereas 11 were diagnosed by surgical biopsy. Two patients were diagnosed in a multidiciplinary meeting with their medical history and radiological findings. All patients were initially treated with methylprednisolone. In five of the cases, steroids had to be switched to azathioprine, methotrexate or mycophenolate mofetil due to side effects or disease progression. Clinical, functional and radiological improvement was observed in 10 cases, whereas 6 remained stable. Mortality rate was 27% (6 patients). Cause of death was respiratory failure in 5 and progression of systemic sclerosis in 1 patient.

Conclusion: NSIP is a challenging disease in both diagnostic and therapeutical aspects. High mortality rates should alarm the clinician in making swift decisions on treatment changes or refering to transplantation.

Keywords: Cryobiopsy, interstistial pneumonia, NSIP