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Title: Autoimmunity profile in patients with combined pulmonary fibrosis and emphysema (CPFE)

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Body: Background: The combination of pulmonary fibrosis and emphysema (CPFE) is a recently defined syndrome, encompassing a distinct radiologic, revealing both upper lobe emphysema and lower lobe fibrosis, as well as lung function profile, with apparently preserved lung volumes contrasting with disproportionally impaired gas exchange,. It has also been recently described in the context of connective tissue diseases. Our primary aim was to investigate the autoimmunity profile of patients with CPFE. Patients and methods: Thirty nine patients, mean age of 66,5 years, 37 men, all smokers, with CPFE based on radiologic and functional criteria (mean FVC: 68,5%pred, FEV1/FVC:78,2, DLCO: 34,9%pred) were recruited on a retrospective and prospective basis. All patients underwent a thorough investigation of their immunologic profile. Results: Fourteen patients (36%) had positive anti-nuclear antibodies (ANA). Patients with positive ANA presented with elevated CD20 levels in lung biopsy specimens suggestive of elevated B cell activity. In additon, 6 patients (15%) presented with positive antineutrophil cytoplasm antibodies (ANCAs) against myeloperoxidase (MPO) indicative of microscopic polyangiitis. Among the latters three developed respiratory and renal insufficiency and were succesfully treated with cyclophosphamide and methylprednisolone whereas the remaining three were switched to azathioprine maintenance treatment.Conclusions: A significant proportion of patients with CPFE may present with underlying auto-immune disorders that may reside incospicuously. Early identification of these patients using a panel of auto-antibodies may lead to more targeted and potentially effective therapeutic applications.