

Differences between ANCA positive and negative lung fibrosis cases without vasculitis

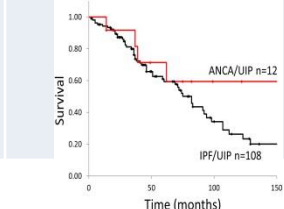
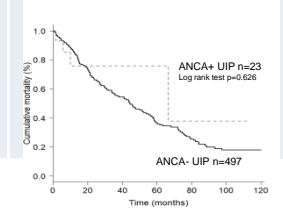
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AIM: Myeloperoxidase anti-neutrophil cytoplasmic antibody is occasionally positive in patients with UIP in the absence of vasculitis. These patients may form a distinct subgroup but are not well characterised. They may have a different prognosis and require different treatment. The aim of this study was to compare ANCA-positive IPF patients without vasculitis with ANCA-negative IPF patients at Ege University Hospital, Izmir, Turkey. We are aware of one similar comparison in Japanese patients (Respirology 2016 21;920-6)

Method: The patients were from Ege University Hospital and submitted to the EMPIRE Registry, which performed the statistical analysis. We compared 23 patients with UIP who were ANCA-positive at presentation but without signs of vasculitis against 497 patients with IPF who were ANCA-negative, comparing demographics, smoking history, comorbidities, HRCT appearances, anti-fibrotics treatment and survival.

Results	ANCA-positive	ANCA-negative	
Number of patients	23	497	
Median age (years)	70	67	p=0.03
Diabetic	4.8%	24%	p=0.02
Pulmonary hypertension	0%	18%	p=0.004
COPD	19%	5%	p=0.024
Definite UIP on HRCT	86%	58%	p=0.007
Lung function tests	No significant differences		
Treated with pirfenidone or nintedanib	48%	57%	
Survival at 72 months	38%	34%	p=0.63

ANCA+ vs ANCA-	Hosuda et al	Ege University
Number of ANCA-positive patients	12 All had surgical biopsies	23
Clinical background	No differences	Less diabetes, less pulmonary hypertension, more COPD
Lung function tests	No differences	No differences
HRCT	More honeycombing surrounded by attenuation	More definite UIP
BAL	More neutrophils	
Histology	More interstitial inflammation More lymphoid follicles More cellular bronchiolitis	
Immunosuppressive treatment	More effective in ANCA+/UIP	
Survival	Tendency to longer in ANCA+/UIP 	Tendency to longer in ANCA+/UIP 

Conclusion: comparing ANCA+ and ANCA- UIP patients

- There are some differences in the clinical background
- The lung function tests show no significant differences
- ANCA+ patients are more likely to have definite UIP on HRCT
- There is a tendency to longer survival for patients who are ANCA+

