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AB0689 EXTRAVASCULAR MANIFESTATIONS OF TAKAYASU ARTERITIS: HISTORICAL COHORT STUDY IN KOREA

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Background: Takayasu arteritis (TAK) is systemic disease characterised by large vessel involvement. Although the vascular characteristics of TAK are well characterised, there is no well-organised study demonstrating the extravascular manifestations of TAK.

Objectives: To evaluate the characteristics of extravascular manifestations of TAK, and to identify the association between vascular and extravascular manifestations of TAK.

Methods: TAK patients from two independent cohorts who fulfilled the 1990 ACR classification and encoded M314 according to ICD-10 code between January 2012 and October 2017 were included in the study. Characteristics of the patients were retrospectively collected from the electronic dataset. A radiologist reviewed CT scans of all included patients to evaluate the pattern of vascular involvement and presence of sacroiliitis. Clinical findings including uveitis, skin lesion, oral ulcer, arthritis, and inflammatory bowel disease (IBD) were reviewed. Logistic regression analysis was performed to evaluate the association between vascular and extravascular manifestation.

Results: A total of 268 TAK patients were included. Mean age at diagnosis was 41.2±14.2 years and 236 (88.1%) were female. The most commonly involved vessel was common carotid artery (176 [65.7%]), and the most common type of vascular involvement was type V (120 [44.8%]). Extravascular manifestation of TAK was observed in 51 (19.0%) patients (table 1). The most common extravascular manifestation was arthritis (axial arthritis [sacroiliitis] [7.1%] and/or peripheral arthritis [6.0%]) (11.9%) followed by recurrent aphthous stomatitis (8.6%) and IBD (2.6%). In multivariable logistic regression analysis, the following factors were significantly associated with presence of arthritis (axial and/or peripheral arthritis): type IIB vascular involvement (adjusted OR 2.956, 95% CI 1.337–6.537, p=0.007) and erythrocyte sedimentation rate (ESR) (adjusted OR 1.014 95% CI 1.003–1.025, p=0.012).

Abstract AB0689 – Table 1. Extravascular manifestations of Takayasu arteritis

	n=268
Any extravascular manifestation	51 (19.0%)
Arthritis (axial arthritis [sacroiliitis] and/or peripheral arthritis)	32 (11.9%)
Recurrent aphthous stomatitis	23 (8.6%)
Inflammatory bowel disease	7 (2.6%)
Erythema nodosum	4 (1.5%)
Uveitis	2 (0.7%)

Conclusions: Extravascular manifestations of TAK are not rare and observed in up to one-fifth of patients. The most common extravascular manifestation was arthritis including sacroiliitis (11.9%). Type IIB vascular involvement pattern and high ESR were significantly associated with arthritis in TAK.

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AB0690 DIAGNOSTIC VALUES OF ENDOTHELIN-1 IN PATIENTS WITH SYSTEMIC NECROTIZING VASCULITIS

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Background: Systemic necrotizing vasculitis (SNV) is characterised by destructive and inflammatory changes in the vessels. Binding of autoantibodies and immune complexes on the surface of endothelial cells stimulates the synthesis of endothelin-1 (ET-1), which leads to activation of macrophages and adhesion of neutrophils, remodelling of the vascular wall and its damage.

Objectives: To evaluate the serum level of ET-1 in patients with SNV and the possibility of its using for the diagnosis of SNV and involvement of individual organs.

Methods: The study included 36 patients with SNV (polyarthritis nodosa – 8, ANCA – associated vasculitis – 28) and healthy controls (n=26). Clinical activities of patients were calculated according to the Birmingham Vasculitis Activity Score (BVAS). All patients had active disease (BVAS >11). The serum levels of ET-1 (pmol/L) were determined by immunoassay analysis using the kits of Biomedica. The outcomes of this study were the differences in marker levels between patients with active SNV and healthy controls, patients with different forms of vasculitis, with varying degrees of BVAS activity, with involvement different organs and systems estimated by analysis of the absolute changes in marker levels and the areas under receiver operating characteristic (ROC) curves (AUC).

Results: The level of ET-1 (M±σ) in the general group of patients with SNV was 0.31±0.24 and did not differ significantly from the control group (0.27±0.10, p>0.05). At the same time, in patients who did not receive at screening glucocorticoids and cytotoxic agents (n=9), it was significantly elevated (0.62±0.58, p=0.03). However, ROC analysis indicated the moderate sensitivity (67%) and the low specificity (48%) of ET-1 for diagnosis of SNV. There were no significant differences in the levels of ET-1 between patients with different forms of vasculitis and with varying degrees of BVAS activity. In the analysis of the values of the ET-1 depending on the involvement of different organs and systems, it was found that only in patients with kidney involvement (n=15) its level (0.40±0.33) was significantly higher compared with patients without kidney involvement (0.28±0.22, p=0.04) and control group (p<0.01). ROC analysis showed that the AUC for ET-1 is 0.75±0.10 (p=0.004), which indicates acceptable capacity for ET-1 differentiate groups of patients with kidney involvement and patients without kidney involvement (sensitivity – 80.0%, specificity – 78.3%).

Conclusions: The serum levels of ET-1 were elevated in patients with SNV with kidney involvement (48% compared to healthy controls and 43% compared with patients without kidney involvement), which can be used for diagnostic purposes.

Disclosure of Interest: None declared

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AB0691 INTERSTITIAL LUNG DISEASE AND MICROSCOPIC POLYANGIITIS IN CHILEAN PATIENTS

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Background: Microscopic Polyangiitis (MPA) is an ANCA associated vasculitis (AAV), associated with p-ANCA (perinuclear) fluorescence pattern and anti-myeloperoxidase (MPO) specificity. Most frequently involved organs are kidney (80%–100%), peripheral nervous system and skin (30%). There is Pulmonary involvement in 25%–35% of patients, being alveolar haemorrhage frequently described. Interstitial lung disease (ILD) has also been recognised.

Objectives: The aim of our study is to report the characteristics of MPA Chilean patients with ILD and to compare it with other series.

Methods: Retrospective study. Patient diagnosed between 2007 and 2016 at the Hospital Clínico Universidad de Chile, with ILD, defined as interstitial lung disease on CT scan with Usual Interstitial Pneumonia (UIP) or Non Specific Interstitial Pneumonia (NSIP) pattern, and MPA were included. Demographic, clinical, laboratory and mortality data were plotted. Data from other series were compared with our results. Other causes that could explain the pulmonary involvement were excluded.

Results: From 94 patients with AAV, 36,1% were MPA, being 16 patients with ILD. All were Hispanic, median age 65.3 years, 32–84 female 62,5% (table 1). Common manifestations were constitutional symptoms (100%), weight loss (68,7%) and fever (68,7%). All patients had anaemia, high ESR (mean 84 mm/hr. range 33–120) and CRP (8–22 times above upper normal limit). All patients were ANCA-p and MPO positive. In 10 cases ILD was diagnosed concomitantly with MPA and in 6 was 0.5 to 15 years before. 4 patients developed pulmonary haemorrhage. Images patterns were 10 UIP and 5 NSIP. All patients received corticosteroid as induction therapy, 15 also received cyclophosphamide. One patient plasmapheresis, and one received Rituximab after a relapsed. Azathioprine was used as