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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

Maintenance therapy. Four patients died during follow-up. table 2 shows data from other worldwide region compared with our data.

Abstract AB0691 – Table 1. Characteristic of chilean patient with ILD an MPA

Age	Sex	CT	Onset (month)	RENAL	MUCOCUTANEOUS	NEUROLOGIC	MSQ	TREATMENT	MORTALITY
55	M	UIP	C	NO	NO	MM	MP/51	CS/CJ/AZA	NO
68	F	UIP	B (10)	NO	LR	PNP	MP/TJ	CS/CJ/AZA	NO
79	M	UIP	B (5)	NO	LR	PNP	MP/51	CS/CJ/AZA	NO
69	F	UIP	C	NO	LR	PNP	SI	CS/CJ/AZA	NO
77	F	UIP	C	GN	LR / CV	MM/NSHL	SI	CS/CJ/AZA	YES
64	F	UIP	C	GN	NO	NO	MP/TJ	CS/CJ/AZA	NO
81	M	UIP	C	GN	NO	NO	NO	CS/CJ/AZA	NO
84	F	UIP/DAH	C	RPGN	Oral Ulcers	NO	NO	CS/CJ/AZA	YES
63	F	UIP	B (168)	GN	NO	CHS/NSHL	NO	CS/CJ/AZA	NO
63	M	NSIP	B (24)	NO	NO	MM	SI	CS/CJ/AZA	NO
53	F	NSIP	C	GN	LR	NO	TJ	CS/CJ/RTX/AZA	YES
68	F	NSIP/DAH	B (50)	NO	NO	PNP	MP	CS/CJ/PLX/AZA	YES
57	M	NSIP/DAH	C	RPGN	NO	PNP	NO	CS/CJ/AZA	NO
78	M	Not UIP not	C	NO	NO	NSHL	MP/51	CS/CJ/AZA	NO
54	F	NSIP	C	NO	LR	PNP	TJ	CS/CJ/AZA	?
32	F	UIP/DAH	B (84)	NO	CV	NO	NO	CS/CJ/AZA	NO

Abstract AB0691 – Table 2. Characteristic of patient with ild and MPA worldwide

Country	CHI	ARG	MEX	CHN	JAP	USA	CAN	PER	IRA
Number of Patient	16	9	19	19	19	3	6	10	40
Age (average)	65.3	58.4	54.2	63.6	66.2	72.3	69.8	70	68
Sex (% female)	62.5	55.5	47.1	58	48	76.7	50	20	39
Onset (M/CA) ILD-MPA	37.5/62.5/0	55.5/54.5/0	82.3/17.7/0	88.4/11.5/0	52.6/47.3/0	33.3/66.7/0	50/50/0	50/50/0	45/45/12
TC (% UIP/NSIP/NON UIP NSIP)	62.5/37.5/25/25	88.8/0/11.2	100/0/0	100/0/0	100/0/0	0/0/100	100/0/0	60/10/30	NA
Mortality %	25	44.4	41.2	31	47	33.3	83.3	50	36

Conclusions: Among chilean patients there are more females, have a more NSIP pattern, and less mortality that other worldwide series.

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AB0692 BELIMUMAB IN COMBINATION WITH AZATHIOPRINE FOR REMISSION MAINTENANCE IN GRANULOMATOSIS WITH POLYANGIITIS AND MICROSCOPIC POLYANGIITIS: EFFECT ON BIOMARKERS

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Background: GPA and MPA (related types of ANCA-associated vasculitis [AAV]) are organ-/life-threatening systemic vasculitides. B cells and increased circulating BLYS (a B cell survival factor) are implicated in AAV pathogenesis, suggesting a role for BLYS in these vasculitides. BEL is an anti-BLYS human immunoglobulin (Ig) G1 λ monoclonal antibody.
Objectives: Examine the effects of BEL plus azathioprine (AZA), on biomarkers in patients with AAV for remission maintenance following standard induction (IND) with glucocorticoids (GC) and CYC or RTX.
Methods: This double-blind, placebo-controlled, multicentre study (BEL115466/ NCT01663623) randomised (1:1) patients (≥ 18 years) with new/relapsing AAV following IND (oral or IV CYC or RTX), to AZA 2 mg/kg/day and oral GC, plus IV BEL 10 mg/kg or PBO (Days 0, 14, 28 and every 28 days until completion). Remission was defined as Birmingham Vasculitis Activity Score=0, plus GC ≤ 10 mg/day. The study was truncated after initiation (n~300 to~100) due to revised AAV standard of care (SoC) affecting recruitment. Biomarker endpoints (serum Igs, B cells and ANCA [anti-MPO/PR3]) were measured at baseline (BL) and thereafter. Summaries by IR were post hoc; no analyses were performed.

Abstract AB0692 – Table 1. Baseline disease characteristics

n (%)	BEL (n=53)	PBO (n=52)
GPA	42 (79)	41 (79)
MPA	11 (21)	11 (21)
ANCA type (historical diagnosis)		
Anti-PR3	41 (77)	40 (77)
Anti-MPO	12 (23)	12 (23)
ANCA positive*	24 (49)	22 (44)
IR		
IV CYC	21 (40)	24 (46)
Oral CYC	18 (34)	15 (29)
Rituximab	14 (26)	13 (25)

ANCA, anti-neutrophil cytoplasm antibodies; BEL, belimumab; CYC, cyclophosphamide; GPA, granulomatosis with polyangiitis; IR, induction regimen; IV, intravenous; MPA, microscopic polyangiitis; MPO, myeloperoxidase; PBO, placebo; PR3, proteinase 3; RTX, rituximab
*n=49 (BEL); n=50 (PBO)

Results: At BL, RTX patients had B cell counts $\leq$ LLQ. CYC patients had notably low BL B cell counts: the lowest were in oral-CYC patients. Circulating memory B cells (CD20+/CD27+) increased rapidly with BEL, then gradually returned to BL (CYC); no major changes occurred with PBO. BEL had no impact on the proportion of naive CD20+CD27– B cells vs PBO, post CYC IND. The number of RTX patients with quantifiable data was low; partial reconstitution occurred in a minority of patients (PBO 2/13; BEL 4/14) and did not translate into vasculitis relapses. Overall Ig levels declined more noticeably with BEL vs PBO; suggesting BEL affects antibody-secreting cells. ANCA+patients were similar between groups (PBO 32/50; BEL 30/49). No trends in change in ANCA status over time occurred, regardless of IR. Individual patient data showed no apparent trends between ANCA titres and AAV activity.

Conclusions: Choice of IR for active AAV affects B cell dynamics. BEL pharmacodynamic effects occurred in patients with AAV receiving SoC. Given the small sample size and high variability, data must be interpreted with caution.

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AB0693 DEMOGRAPHIC, CLINICAL, LABORATORY AND IMAGING CHARACTERISTICS OF AN INCIDENCE COHORT OF 93 PATIENTS WITH LARGE VESSEL GCA

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Background: Few study have evaluated the clinical and imaging data of patients with large vessel GCA (LVGCA)
Objectives: To evaluate clinical, demographical and imaging data of an inception cohort of 93 patients with LVGCA recruited in Reggio Emilia (Northern Italy)
Methods: All patients with incident large vessel GCA diagnosed between 1 January 2005 and 31 December 2016 and living in the Reggio Emilia area, were identified by capture and re-capture checking of computerised discharge diagnosis codes (ICD10) and using outpatients databases from rheumatology, internal medicine, surgery, pathology, imaging departments of Reggio Emilia Hospital as well