FRI0193

ULTRASONOGRAPHY IN THE DIAGNOSIS OF GIANT CELL ARTERITIS

R. Jese¹, Z. Rotar¹, M. Tomsic^{1,2}, A. Hocevar^{1,2} on behalf of: ¹University Medical Centre Ljubljana, Department of Rheumatology, Ljubljana, Slovenia; ²Faculty of Medicine, University of Ljubljana, Ljubljana, Slovenia

Background:

Objectives: To evaluate the frequency of cranial and aortic arch artery involvement in GCA using color Doppler ultrasonography (CDS).

Methods: We performed CDS of cranial and aortic arch arteries in 248 incipient, clinically diagnosed, GCA patients (64.9% females, median (IQR) age 75 (67-80) years) between October 2013 and September 2019, using a Philips IU22 with 5–17.5 MHz linear probe or Philips Epiq 7 with 5–18.5 MHz linear probe. Temporal, facial, occipital, carotid, vertebral, subclavian, and axillary arteries were examined bilaterally. A halo with positive compression sign was considered a positive finding. Additionally, the thickness of intima-media complex (IMT) of individual vessel was measured, and compared to the IMT of 97 consecutive suspected GCA cases (60.8% females median (IQR) age 74 (65-81) years), in whom GCA was excluded, that served as a control group.

Results: The CDS was positive in 244 (98.4%) patients in at least one of the examined arteries. Temporal arteries were most commonly affected, and were involved in 192 (77.4%) patients, followed by facial and occipital arteries, involved in 122 (49.2) and 72 (29.0%) patients, respectively. Extracranial large vessel involvement (LVV) was found in 87 (35.1%) patients (32 patients had isolated LVV, and 55 concomitant cranial and LVV artery involvement). Among the 161 patients without LVV, 12 (4.8% of the studied cohort) had involvement of cranial arteries other than temporal arteries (we found facial and occipital artery involvement in 11 and 3 patients, respectively). Table 1 shows the frequency of individual vessel involvement in GCA, and the IMT of CDS inflamed and non-inflamed arteries in GCA, and in controls.

Table 1. The involvement of cranial and aortic arch arteries in GCA assessed by CDS and intima-media thickness of inflamed and non-inflamed arteries in GCA, and controls

Artery	GCA No (%)	IMT (mm) in GCA		IMT (mm) in Controls
	Positive CDS	Positive CDS*; minimal	Negative CDS	Negative CDS; maximal
Temporal	192 (77.4)	0.71±0.19; 0.33	0.25±0.07	0.23±0.05; 0.46
Facial	122 (49.2)	0.75±0.27; 0.41	0.29±0.07	0.26±0.07; 0.47
Occipital	72 (29.0)	0.73±0.33; 0.45	0.26±0.06	0.23±0.05; 0.46
Carotid	34 (13.7)	1.53±0.44; 0.88	0.78±0.18	0.72±0.15; 1.09
Vertebral	25 (10.1)	1.33±0.47; 0.74	0.45±0.10	0.42±0.08; 0.63
Subclavian	67 (27.0)	1.65±0.45; 0.91	0.70±0.14	0.70±0.13; 0.99
Axillary	59 (23.8)	1.74±0.65; 1.00	0.61±0.17	0.57±0.13; 0.97
Any artery	244 (98.4)	-	-	-

Legend: GCA giant cell arteritis; IMT thickness of intima-media complex; * mean±SD;

Conclusion: CDS of seven preselected cranial and aortic arch arteries provides a high diagnostic yield in GCA.

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IDENTIFYING PHENOTYPES OF PATIENTS WITH BEHÇET'S SYNDROME BY A CLUSTER ANALYSIS IN A LARGE COHORT

J. Zou¹, J. Guan¹. ¹Huadong Hospital, Fudan University, Division of Rheumatology, Shanghai, China

Background: Behçet's syndrome (BS) is a clinical entity with heterogeneous clinical manifestations (1). The symptoms and major organ involvement of BS tend to vary among gender and age (2).

Objectives: To investigate the gender associated clinical features and aggregate of patients with BS based on phenotypes and organ involvement in a well-defined large cohort of patients with BS.

Methods: This was a single-center study of 1,596 consecutive BS patients in Shanghai, China from 2012 September to 2019 December. We compared clinical variables and conducted a hierarchical cluster analysis applied according to twenty-five clinical variables to determine subgroups of patients.

Results: A total of 850 BS patients were included. Male gender was associated with ocular involvement (RR 2.33), vascular involvement (RR 2.05) and cardiac lesions (RR 5.52). Female gender associated with genital ulcers (RR 0.85). Four clusters (C1–C4) were yielded. Cluster C1 (n=165, 19.9%), merely

oral and genital ulcer type: consisted predominantly of older females. Disease severity was relatively low, Krause score = 2.32. In C2 (n=365, 43.9%), mix type: was an equal gender distribution with older onset age. Most patients had erythematous nodosum (64.1%) and genital ulcers (82.2%). Cardiac and vascular involvement were observed in 9.3% and 12.1%, which were the highest frequencies among the four clusters. C3 (n=160, 19.3%), intestinal type: was an equal sex ratio with older onset age. All patients presented with intestinal ulcer, while fewer patients had erythematous nodosum and pseudofolliculitis, 20.0% and 25.0% respectively. In C4 (n=141, 17.0%), uveitis type: was predominantly male with younger onset age. Intestinal involvement was negatively associated with uveitis [0.24, (0.13, 0.46), $P < 0.0001$]. No patients had cardiac involvement in C3 or C4. Regarding the sensitivity of classification criteria, Cheng and Zhang (China) remain the highest in all clusters compared with Japan revised criteria, ICBD, ISG.

Conclusion: From our large cohort, the gender-phenotypes analyses suggest that male is a risk factor for ocular and cardiovascular in BS patients. We identify four unique subphenotypes representing different clinical features, intestinal and ocular phenotype clusters represent the two most common manifestations of major organ involvement in BS.

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TABLE 1. Characteristics of patients with Behçet's syndrome after clustering on clinical manifestations

Characteristics	C1 (n=165)	C2 (n=356)	C3 (n=160)	C4 (n=141)
Age at onset (IQR), years	29 (20-40)	28 (20-35)	27 (20-38)	25 (19-32)
Disease duration (IQR), years	5 (3-10)	8 (3-10)	6 (3-10)	7 (4-10)
Male gender (%)	38.9	52.3	51.2	75.2
Classification criteria				
JPN (%)	0	69.6	25.0	83.7
CHN (%)	98.8	96.7	97.5	99.3
ICBD (%)	98.8	89.9	65.6	99.3
ISG (%)	0.6	72.1	25.0	81.6
Oral ulceration (%)	100	100	98.1	99.3
Genital ulceration (%)	98.2	82.2	63.1	58.9
Erythema nodosum (%)	0	64.1	20.0	48.9
Pseudofolliculitis (%)	0	34.5	25.0	27.0
Arthritis/Arthralgia (%)	0	36.2	11.6	14.2
Uveitis (%)	0.6	0.8	1.9	100
Gastrointestinal involvement (%)	0	1.1	100	2.7
Vascular involvement (%)	2.4	12.1	1.2	4.3
Cardiac lesion	3.6	9.3	0	0
Central nerve system involvement (%)	4.2	3.8	2.5	3.5

JPN: Japan revised criteria; CHN: Cheng and Zhang (China) criteria; ICBD: International Criteria for Behçet's disease; ISG: International Study Group for Behçet's disease.

Disclosure of Interests: None declared

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ULTRASONOGRAPHIC HALO SCORE ASSOCIATES WITH INTIMAL HYPERPLASIA IN GIANT CELL ARTERITIS

K. Van der Geest^{1,2}, F. Borg¹, K. Wolfe¹, W. A. Schmidt³, R. Luqmani⁴, B. Dasgupta¹. ¹Southend University Hospital, Westcliff-on-Sea, United Kingdom; ²University Medical Center Groningen, Groningen, Netherlands; ³Immanuel-Krankenhaus GmbH, Berlin, Germany; ⁴University of Oxford, Oxford, United Kingdom

Background: We have recently developed a novel ultrasonographic Halo Score to quantify the extent of vascular inflammation in GCA [1]. High Halo Scores were associated with a high rate of ocular ischaemia among patients with GCA. Earlier studies have shown that GCA patients with intimal hyperplasia in their temporal artery biopsy (TAB) are at the highest risk of neuro-ophthalmic, ischaemic complications [2,3]. We therefore questioned whether the ultrasonographic Halo Score might be linked to the presence of intimal hyperplasia in patients with GCA.

Objectives: To investigate the relationship between the ultrasonographic Halo Score and intimal hyperplasia.

Methods: This is a prospective study including 92 patients suspected of having GCA, who underwent both ultrasound of temporal/axillary arteries and TAB at diagnosis. Ultrasonographic halo counts and Halo Scores were determined [1]. An experienced pathologist determined whether or not the TAB findings were consistent with GCA. TABs were systematically evaluated for the presence of a transmural infiltrate and intimal hyperplasia. Multiple linear regression analysis was performed with either halo counts or Halo Scores as the dependent variable.

Predictive variables included the presence of a transmural TAB infiltrate, intimal hyperplasia and male sex.

Results: The TAB was consistent with GCA in 27 patients. The TAB revealed transmural inflammation in 18 patients and giant cells in 24 patients. Intimal hyperplasia was found in 20 patients with a positive TAB. Patients with a positive TAB showed higher halo counts and Halo Scores than patients with a negative TAB. Overall, patients with a positive TAB and intimal hyperplasia presented with the highest halo counts and Halo Scores (Figure). Among patients with a positive TAB, only intimal hyperplasia and male sex were predictive of higher halo counts and Halo Scores in the multiple linear regression analysis. Ocular ischaemia was present in 14% of patients with a positive TAB without intimal hyperplasia. However, 40% of patients with a positive TAB and intimal hyperplasia suffered from ocular ischaemia.

Conclusion: The ultrasonographic Halo Score is strongly influenced by the presence of intimal hyperplasia, a TAB feature that associates with cranial ischemic complications in patients with GCA [2,3].

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Figure

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INVOLVEMENT OF THE PULMONARY ARTERIES IN PATIENTS WITH TAKAYASU ARTERITIS

X. Kong¹, L. MA¹, P. LV¹, X. Cui¹, R. Chen¹, Z. Ji¹, H. Chen¹, J. Lin¹, L. Jiang¹.

¹Zhongshan Hospital affiliated to Fudan University, Shanghai, China

Background: Takayasu arteritis (TA) is a chronic, granulomatous large-vessel vasculitis. It involves the aorta and its main branches predominantly, and leads to vascular thickness, stenosis and occlusion [1]. Besides the aorta and its branches, pulmonary arteries (PAs) are involved in TA. PAs have been reported to be involved in 6.9% to 80% of TA patients from different populations [2-3].

Objectives: We investigated the clinical characteristics, pulmonary parenchymal features and cardiac functions in TA patients with PA involvement by combining multiple imaging modalities (MRA, CTA, PET-CT, lung VQ scan, echocardiography and high-resolution computed tomography (HRCT)). Our aim was to elicit

better understanding of TA patients with PA involvement to aid rational treatment for these patients and improve their prognosis.

Methods: We enrolled 216 patients with TA from a large prospective cohort. PAI was assessed in each patient based on data from magnetic resonance angiography/computed tomography angiography. Pulmonary hypertension, cardiac function, and pulmonary parenchymal abnormalities were evaluated further in patients with PAI based on echocardiography, New York Heart Association Functional Classification and pulmonary computed tomography, respectively. These abnormalities related to PAI were followed up to evaluate treatment effects.

Results: PAI was detected in 56/216 (25.93%) patients, which involved the pulmonary trunk, main PAs and small vessels in the lungs. Among patients with PAI, 28 (50%) patients were accompanied by pulmonary hypertension, which was graded as 'severe' in 9 (16.07%), 'moderate' in 10 (17.86%) and mild in 9 (16.07%). Forty (71.43%) patients had cardiac insufficiency (IV: 6, 10.71%; III: 20, 35.71%; II: 14, 25.00%). Furthermore, 21 (37.50%) patients presented with abnormal parenchymal features in the area corresponding to PAI (e.g., the mosaic sign, infarction, bronchiectasis). During follow-up, two patients died due to abrupt pulmonary thrombosis. In the remaining patients, the abnormalities mentioned above improved partially after routine treatment.

Conclusion: PA involvement is very common in TA patients. Physicians should be alerted to PA involvement even if obvious pulmonary symptoms are absent because they can cause PH, cardiac insufficiency as well as pulmonary parenchymal lesions, which will worsen the prognosis.

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Figure 1. Imaging of PA lesions in TA patientsA: Dilation of the pulmonary trunk; B: thickening of the pulmonary trunk; C: stenosis of the right main PA; D: embolism of lower PAs on both sides; E: inflammation of the pulmonary-trunk root upon PET-CT; F: absence of left PAs and stenosis of the right main PA; G-I: pulmonary MRA (G), CTA (H) and VQ scan (I) of a patient with TA. MRA shows a fine right main PA and low perfusion in the right lung (G); CTA demonstrates a fine right main PA and fewer PA branches in the right lung (H); lung VQ scan shows multiple arterial emboli in the right lung and obvious less blood supply to the right lung.

Figure 2. Pulmonary lesions on HRCT. A: The mosaic sign in the left lung; B: Pulmonary infarction of the right middle lobe; C: Mild pleural effusion on the left side; D: Bronchiectasis in the right lung; E-F: Ground-glass opacity (E) in the right upper lobe of a TA patient with an embolism of the right upper pulmonary branches (F); G-I: Cavitation (G) and mass-like consolidation (H) in the patient with severe stenosis of right main pulmonary artery (I).