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ULTRASONOGRAPHY IN THE DIAGNOSIS OF GIANT CELL ARTERITIS

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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

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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.

References:

- [1] Bettiol A, Hatemi G, Vannozzi L, et al. Treating the Different Phenotypes of Behçet's Syndrome. *Front Immunol* 2019;10:2830.
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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.

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ULTRASONOGRAPHIC HALO SCORE ASSOCIATES WITH INTIMAL HYPERPLASIA IN GIANT CELL ARTERITIS

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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.