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THU0298 INITIAL MANIFESTATIONS AND OUTCOME OF PATIENTS WITH INCOMPLETE BEHÇET'S SYNDROME

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Background: Behçet's syndrome (BS) has a heterogeneous expression involving many organ systems and is diagnosed by recognizing the coexisting manifestations. Several patients initially have few of these manifestations and do not fulfill the International Study Group (ISG) criteria. When these are major organ manifestations, failure to recognize BS and treat promptly may lead to permanent damage in these organs.

Objectives: The aim of this study is to highlight the magnitude of this problem by surveying the frequency, presentation patterns and outcome of patients who did not fulfill ISG criteria when they presented to our clinic, but were followed and treated for manifestations strongly suggesting BS.

Methods: We conducted a retrospective chart review of all BS patients who were registered between 2003 and 2008. Among these 2385 patients, 199 (8%) BS patients who did not fulfill ISG criteria at their initial visit were included in this study. Patients were called and a standard form was used for collecting demographic characteristics, BS manifestations at initial visit and during follow-up and treatment.

Results: Among the 199 patients (M/W: 90/109, mean age: 34 ± 11 years) who did not fulfill ISG criteria when they presented to our clinic, 70 (35%) had major organ involvement. The types of major organ involvement that led to a diagnosis of BS at initial visit despite not fulfilling ISG criteria were eye involvement in 37, vascular involvement in 29 (venous thrombosis in 22, arterial aneurysms in 7), nervous system involvement in 3 and gastrointestinal (GI) involvement in 1 patient. Thirty-five patients (18%) had a family history of BS. Of 199 patients, 167 had at least one more visit with a median follow-up of 11 years (IQR: 7-12). We were able to contact 116 of these patients and saw that 53 had fulfilled ISG criteria in the meantime. Among the 51 patients that we were not able to contact, 17 had fulfilled criteria while they were being followed in our clinic. Thus, a total of 70 (42%) patients fulfilled ISG criteria after a median follow-up of 1.5 years (IQR: 1-4.25). All but 2 patients who developed eye involvement during the follow-up had fulfilled ISG criteria with a new mucocutaneous manifestation. After a median follow-up of 4 years (IQR: 1-7), 23 (14%) patients had developed at least one non-criteria BS manifestation, including vascular involvement in 10, arthritis in 13, neurologic involvement in 2 patients and GI involvement in 1 patient.

Among the 81 patients who developed at least one new manifestation, 16 (20%) were under immunosuppressive or interferon-alpha treatment at the time they developed their new manifestation. The remaining 65 patients had only received colchicine.

Among the 29 patients who had vascular involvement at initial visit, 7 (24%) had a vascular relapse at different vascular site.

Three patients had another rheumatologic diagnosis (ankylosing spondylitis, seropositive rheumatoid arthritis and sarcoidosis) at the end of the follow-up.

Conclusion: In our 10-year follow-up cohort study, 42% of the 167 incomplete BS patients had fulfilled ISG criteria within a median duration of 1.5 years. Sixty-two (37%) of these had major organ involvement that could have caused severe morbidity and mortality if the diagnosis of BS was missed and patients were untreated.

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THU0299 PATIENT GLOBAL ASSESSMENT OF DISEASE ACTIVITY IN BEHÇET'S SYNDROME: A MULTICENTER STUDY

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Background: Disease activity evaluation is mandatory according to the OMERACT Core Set of Domains for Behçet's Syndrome (BS). Poor data are available on Patient (PtGA) and physician (PGA) global assessment of disease activity in BS.

Objectives: To assess PtGA performance in patients with BS and how different disease manifestations influence the patient and physician's perception of disease activity.

Methods: A multicenter cross-sectional cohort of consecutive BS patients was enrolled. Disease activity was evaluated by PtGA, PGA and the Behçet's disease current activity form (BDCAF). PtGA and PGA were assessed through a single question ("How active was BS during the last week?") in a 10-cm visual analogic scale. Health related quality of life (HRQoL) perception was evaluated by the Physical Component Summary (PCS) and the Mental Component summary (MCS) of the SF36v2 questionnaire.

Correlation between PtGA, PGA, BDCAF and between them and SF36v2 component summaries was assessed through the Spearman's correlation coefficient (rho). Two separated multiple regression models were built to measure the independent effect (β regression) of each active clinical manifestation on PtGA and PGA.

Results: Overall, 226 BS patients from 5 Mediterranean Countries were enrolled. Out of them, 111 (49.1%) were males and the median (IQR) age and disease duration were 46.9 (35.6-55.2) and 11.7 (5.9-20.8) years, respectively.

The median (IQR) value of PtGA, PGA and BDCAF were 2.0 (0.3-5.0), 1.0 (0.0-3.0) and 3.0 (0.0-5.0), respectively. PtGA significantly correlated (Figure) with both PGA (rho 0.759. p < 0.001) and BDCAF (rho 0.523. p < 0.001), in support of its construct validity. PtGA scores were significantly higher than PGA with a mean (SD) difference of 0.8 (1.8), suggesting a different weighing of disease activity between patients and physicians. Indeed, PtGA and PGA were differently influenced by active clinical manifestations (Table). PtGA was mainly dependent on active mucocutaneous (β 0.243), gastrointestinal (β 0.216) and ocular (β 0.209) involvement whereas a major effect of ocular manifestations was observed in PGA (β 0.378). PtGA was correlated with worse HRQoL perception measured by PCS (rho -0.476. p > 0.001) and MCS (rho -0.451. p > 0.001).

Conclusion: PtGA seems to be a valid outcome measure in BS and should be considered as a separate outcome measure or better be included in composite index for overall disease activity evaluation.

Abstract THU299 – Table 1.

PtGA	PGA
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Active involvement	β	p	Active involvement	β	p
Mucocutaneous	0.243	<0.001	Ocular	0.378	<0.001
Gastrointestinal	0.216	0.001	Mucocutaneous	0.284	<0.001
Ocular	0.209	0.001	Gastrointestinal	0.192	<0.002
Vascular	0.108	0.086	Vascular	0.129	<0.027
Arthritis	0.088	0.185	Arthritis	0.121	0.048
Neurologic	0.003	0.962	Neurologic	0.047	0.420

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Figure. Correlation between PtGA and other outcomes measures

PtGA: patient's global assessment, PGA: physician's global assessment, BDCAF: Behçet's disease current activity form, PCS: physical component summary; MCS: mental component summary (MCS)

Abstract THU0299 – Figure 1

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THU0300

IMPACT OF VASCULAR ULTRASOUND ON EVALUATION OF GIANT CELL ARTERITIS AMONG 503 PATIENTS IN AN ACADEMIC MEDICAL CENTER

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Background: As vascular ultrasound (VUS) is increasingly used in evaluation of giant cell arteritis (GCA), its impact on clinical care is of great interest.

Objectives: To describe utilization of VUS in GCA evaluation at a large academic medical center and investigate impact of VUS on clinical impression.

Methods: We performed a retrospective cohort study of patients who underwent VUS at a single center in Boston, 2013-2017, to evaluate suspected or known GCA. Trained cardiovascular ultrasound technicians used a standardized protocol to image the temporal, carotid, subclavian, and axillary arteries for presence of hypoechoic circumferential wall thickening (halo sign), hyperechoic wall thickening, stenosis, and occlusion. VUS images were interpreted by trained cardiovascular medicine physicians as consistent with acute arteritis ("acute"), no evidence of arteritis ("none"), or hyperechoic wall thickening without halo sign ("hyperechoic"). Demographic, laboratory, medication, pathology and clinical data including the treating physician's clinical suspicion for GCA pre- and post-VUS were obtained by electronic medical record review. Fisher's exact test and Wilcoxon rank-sum tests compared baseline characteristics among patients with VUS positive for acute arteritis ("acute") vs. negative for acute arteritis ("none" or "hyperechoic"). We compared the treating physician's pre- and post-VUS clinical suspicion for GCA among patients with no history of GCA or aortitis.

Results: We identified 503 patients with median age 70.4 years; 69.0% were female and 87.5% White. VUS interpretation was acute in 48 patients (9.5%), none in 427 (84.9%), and hyperechoic in 28 (5.6%). Baseline characteristics are presented in Table 1. Weight loss, cranial symptoms, higher ESR, and steroid use were more common in patients with VUS positive for acute arteritis. Change in the treating physician's clinical suspicion for GCA is shown in Figure 1. VUS positive for acute arteritis increased suspicion for GCA in 26/35 (74%) cases, and VUS negative for acute arteritis lowered suspicion for GCA in 303/396 (77%). Of 110 patients with a temporal artery biopsy after VUS, 16 (14.6%) biopsies showed active arteritis; 11 of these were in patients with VUS negative for acute arteritis.

Conclusion: VUS result changed the clinical suspicion for GCA in approximately 75% of patients in this large cohort.

Abstract THU300 – Table 1. Characteristics at the time of VUS

Age, years	70.4 (62.7, 77.5)	73.2 (65.8, 79.7)	69.9 (62.6, 77.3)	NS
History of GCA or aortitis	14.3	27.1	13.0	0.02
History of PMR	31.2	22.9	32.1	NS
New headache	51.3	60.4	50.3	NS
Weight loss	13.3	22.9	12.3	0.05**
Fatigue	36.2	45.8	35.2	NS
Jaw claudication	10.9	33.3	8.6	<0.01
Scalp tenderness	17.1	37.5	15.0	<0.01
TA tenderness/decreased TA pulse	14.5	29.2	13.0	<0.01
Transient vision loss	7.8	10.4	7.5	NS
Permanent vision loss	3.0	6.3	2.6	NS
CRP*, mg/L	15.7 (3.1, 76.0)	36.3 (8.1, 81.9)	13.7 (2.9, 75.3)	NS
ESR*, mm/h	40 (18, 83)	67 (30, 95)	40 (17, 81)	<0.01
Steroids at time of VUS	57.5	75.0	55.6	0.01 0.02
>0 to <15mg/day	34.3	30.6	34.8	NS
≥15 to <40mg/day	23.5	8.3	25.7	
≥40mg/day	42.2	61.1	39.5	
Steroids ≥3 weeks	56.8	41.7	59.3	

Presented as% or median (interquartile range).

CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; GCA: giant cell arteritis; PMR: polymyalgia rheumatica; TA: temporal artery; VUS: vascular ultrasound. NS: not significant.

*Among 418 patients with CRP and 370 with ESR checked within 3 weeks before VUS

**p=0.046