Table 1. Baseline characteristics of GCA patients and controls in our retrospective cohort. *CDUS=Colour Doppler Ultrasound; **Temporal Artery Biopsy; ***18-FDG-Positron Emission Tomography/Computed Tomography.

	GCA patients (n=62)		Controls (n=144)	
	Range		Range	
Gender, % female (n)	66.1 (41)		50.7 (73)	
Age (years), mean (SD)	74.2 (9.4)	51 – 91	70.2 (11.0)	50 – 97
ESR, mean (SD)	78.4 (30.3)	22 – 120	39.9 (32.4)	2 – 124
Missing	3		9	
CRP, mean (SD)	74.7 (60.0)	2 – 283	35.8 (52.9)	0.4 – 280
Missing	1		6	
Polymyalgia Rheumatica, % yes (n)	21 (13)		11.8 (17)	
Cranial symptoms, % yes (n)	80.6 (50)		75.0 (108)	
Vision loss, %yes (n)	27.4 (17)		15.3 (22)	
Compliant with GCA (i.e. AION, central retinal artery occlusion diplopia)				
Diagnostics				
CDUS* %yes (n)	96.8 (60)		96.5 (139)	
TAB** %yes (n)	58.1 (36)		18.8 (27)	
PET/CT***yes (n)	40.3 (25)		16.0 (23)	

Figure 1. Description of consultation (A); referral (B); diagnostic (C) and total (D) delay in weeks for controls (black) and GCA patients (green), stratified for C-GCA (red), LV-GCA (blue) and C/LV-GCA (purple). ¹Not possible for controls as no GCA-indicated treatment was started.

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AB0368

CLINICAL CHARACTERISTICS AND RADIOGRAPHIC OUTCOME OF VASCULAR BEHCET'S DISEASE INVOLVING AORTA AND ITS MAJOR BRANCHES

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Background: Behcet's disease (BD) is an immune-mediated systemic vasculitis affecting any size of blood vessels. Previous studies showed that BD may involve large arterial vessels [1] but the radiographic outcome has not been well known due to its low prevalence.

Objectives: To evaluate the radiographic outcome of vascular BD involving aorta and its major branches and the clinical characteristics of each prognosis group.

Methods: We retrospectively analyzed the medical records of patients who visited a single-center tertiary hospital in Seoul, Korea, across a 19-year period (Jan 2002 to Dec 2020). All patients fulfilled the International Criteria for Behcet's Disease. The patients included in this study had undergone CT angiography (CTA) of the aorta and its major branches at least twice in a 2–5 year interval and showed characteristic vascular involvements (aneurysm, stenosis, occlusion, vessel wall thickening, and thrombosis). Due to lack of the validated definition of radiographic progression of large vessel vasculitis in BD, we defined 20% changes of existing lesions or occurrence of new lesions as a radiographic progression.

Results: 22 patients with BD met the inclusion criteria. 5 patients (22.7%) progressed radiographically between the initial and follow-up CTA despite low acute

phase reactant levels (AUC of ESR and CRP were 13.94 mm/h and 0.32 mg/dl, respectively). Aneurysm was the most frequent form of vascular involvement followed by thrombosis (21 [95.5%] and 11 [50.0%] patients, respectively, Table 1). Patients with radiographically progressive disease had longer disease duration (1.63 vs. 6.61 years, $p=0.010$) at initial CTA evaluation. Other clinical characteristics, including ESR, CRP, medication use, and follow up duration, were not different between two groups (Table 2).

Table 1. Radiographic characteristics of the cohort.

	Overall	Not progressed	Progressed	
n	22	17	5	p
Aneurysm (%)	21 (95.5)	16 (94.1)	5 (100.0)	>0.999
Stenosis (%)	5 (22.7)	4 (23.5)	1 (20.0)	>0.999
Occlusion (%)	5 (22.7)	5 (29.4)	0 (0.0)	0.440
Wall thickening (%)	4 (18.2)	4 (23.5)	0 (0.0)	0.589
Thrombosis (%)	11 (50.0)	9 (52.9)	2 (40.0)	>0.999

Conclusion: A considerable number of vascular BD patients involving aorta and its major branches progressed radiographically despite low disease activity. Radiographically progressed patients had longer disease duration, but other clinical characteristics were not significantly different whether progressed or not.

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Table 2. Clinical characteristics of the cohort.

	Overall	Not progressed	Progressed	
n	22	17	5	p
Sex (Male) (%)	22 (100.0)	17 (100.0)	5 (100.0)	NA
Age (yr)	46.43 (7.71)	45.06 (8.06)	51.10 (4.03)	0.126
Disease duration (yr)	2.77 (3.99)	1.63 (2.66)	6.61 (5.61)	0.01
HTN (%)	5 (22.7)	4 (23.5)	1 (20.0)	>0.999
DM (%)	1 (4.5)	1 (5.9)	0 (0.0)	>0.999
Dyslipidemia (%)	6 (27.3)	5 (29.4)	1 (20.0)	>0.999
CKD (%)	7 (31.8)	4 (23.5)	3 (60.0)	0.321
BMI (kg/m ²)	42.91 (91.28)	48.23 (103.94)	24.82 (1.69)	0.626
AUC of CRP (mg/dl)	0.32 (0.37)	0.31 (0.41)	0.37 (0.12)	0.758
AUC of ESR (mm/h)	13.94 (11.40)	13.30 (12.77)	16.12 (4.66)	0.638
Immunosuppressive agent use (%)	18 (81.8)	14 (82.4)	4 (80.0)	>0.999
Steroid use (%)	22 (100.0)	17 (100.0)	5 (100.0)	NA
f/u duration (yr)	3.54 (0.80)	3.56 (0.79)	3.50 (0.92)	0.885

Data are presented as mean (SD) unless otherwise specified

Disclosure of Interests: None declared

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AB0369

EVALUATION OF BASELINE POSITRON EMISSION TOMOGRAPHY IN THE DIAGNOSIS AND ASSESSMENT OF GIANT CELL ARTERITIS

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Background: Positron emission tomography (PET/CT) has gained importance in the diagnosis and assessment of large vessel vasculitis (LVV) recently.

Objectives: We aimed to investigate the diagnostic importance and clinical significance of PET/CT findings in giant cell arteritis (GCA).

Methods: Data of the patients who underwent PET/CT to investigate large vessel involvement and who had at least 6 months of follow-up with a clinical diagnosis of GCA were retrospectively evaluated. PET/CT images were assessed by an experienced nuclear medicine specialist, regions of interest were drawn for major vascular territories and standardized maximum uptake values (SUVmax) of these areas were recorded.

Results: Twenty-nine consecutive patients (median age 68 (50-83), mean follow-up time 37.1 ± 48.8 (6-242)) were included into the study. All patients were over 50 years old and had erythrocyte sedimentation rate (ESR) over 50 mm/h at the time of imaging. Twenty patients (68.9%) met the ACR 1990 Classification criteria (ACR (+) group). The number of patients who had hypermetabolism in the aorta and its major branches in favour of LVV in PET/CT was 23 (79.3%) (PET-CT (+) group). Thoracic and abdominal aorta involvement were detected in 22 (75.8%) and 16 (55.2%) patients, respectively. There was positive correlation

between SUVmax in thoracic and abdominal aorta on PET/CT and ESR at diagnosis ($r = 0.63$ $p = 0.002$ and $r = 0.77$ $p < 0.001$, respectively) and SUVmax in thoracic aorta and CRP ($r = 0.50$ $p = 0.026$). PET/CT (-) patients had more frequent disease flares during the follow-up (4/6 vs. 5/23 $p = 0.035$ OR = 7.2 (1.01–51)). Three distinct subgroups were defined by implementing both ACR criteria and PET/CT positivity. Among ACR (+) patients ($n=20$); comparison of PET/CT (+) ($n=14$) and PET/CT (-) ($n=6$) patients did not show any difference in age at diagnosis, presence of polymyalgia rheumatica (PMR), flare rate and damage scores. Among PET/CT (+) patients ($n=23$), the mean age at diagnosis was higher, PMR and bilateral axillary artery involvement was more frequent in ACR (+) group ($n=14$) (Table 1).

Conclusion: PET/CT is increasingly used in the diagnosis and assessment of GCA in our center. The level of FDG uptake of the vessel wall in PET/CT correlates with the acute phase response. Flare was rarely observed in PET/CT (+) patients at diagnosis. Axillary artery involvement detected on PET/CT may be associated with the classical GCA clinic in ACR(+) patients (1). PET/CT (+) patients who does not met ACR criteria seems to have a diverse clinic features like young age and rare presence of PMR. PET/CT findings may be helpful in recognizing subgroups and predicting prognosis of GCA although prospective studies with follow-up scans are warranted.

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Table 1. Comparison of patients who fulfilled and not fulfilled ACR 1990 classification criteria among PET/CT (+) patients.

	ACR (+) PET/CT (+) (n=14)	ACR (-) PET/CT (+) (n=9)	p	OR (%95 CI)
Age at diagnosis	68.8±4.5	63.3±9.2	0.004	
PMR	10	2	0.021	2.5 (1 – 6.1)
History of flare	4	1	NS	
CRP at diagnosis	75.1±30.6	130.8±93.4	0.024	
ESR at diagnosis	93.9±28.1	112.5±21.2	NS	
Brachiocephalic artery	9	6	NS	
Right subclavian	8	5	NS	
Left subclavian	9	5	NS	
Right carotid	8	5	NS	
Left carotid	9	6	NS	
Right axillary	7	0	0.011	2 (1.18 – 3.3)
Bilateral axillary	6	0	0.022	1.75 (1.1-2.7)
Thoracic aorta SUVmax (mean)	3.9±1.1	4.6±1.3	NS	
Abdominal aorta SUVmax (mean)	4.5±1.2	5.3±1.8	NS	

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AB0370

UTILITY OF CRP AND ESR IN THE DIAGNOSIS OF GIANT CELL ARTERITIS RELAPSE IN A PHASE 2 TRIAL OF MAVRILIMUMAB

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Background: No universally accepted definition of flare currently exists in giant cell arteritis (GCA). Although relapses are defined mostly on clinical grounds (recurrence of GCA-related signs/symptoms), C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) help clinicians assess disease activity. In fact, >70% of patients on glucocorticoids (GCs) alone have increased CRP or ESR when the disease is active. In contrast, tocilizumab, given its IL-6-blockade effect in the liver, rapidly reduces CRP and ESR levels, rendering them unreliable for disease activity monitoring. Mavrilimumab – a GM-CSF receptor α inhibitor with demonstrated efficacy in a Phase 2 GCA trial¹ – downregulates inflammation upstream of IL-6. We hypothesized that mavrilimumab would not interfere with the utility of CRP and ESR in monitoring disease activity and in identifying GCA relapse.

Objectives: To analyze the relationship between CRP/ESR and clinical disease activity in GCA patients treated with mavrilimumab.

Methods: New-onset and relapsing GCA patients with active disease were recruited. GC-induced remission (no GCA symptoms and CRP <1 mg/dL or ESR <20 mm/hr) was required by baseline. Patients were randomized 3:2 to mavrilimumab 150 mg or placebo subcutaneously every 2 weeks plus a protocol-defined 26-week prednisone taper. The primary efficacy endpoint was time to relapse by Week 26. Relapse (adjudicated) was defined as recurrent GCA-related signs/symptoms, including new/worsening vasculitis on imaging, concurrent with CRP ≥ 1 mg/dL and/or ESR ≥ 30 mm/hr. CRP and ESR were also measured periodically during the trial.

This post hoc analysis assessed the association of recurrent GCA-related signs/symptoms with concurrent CRP or ESR elevation post-randomization by treatment arm. We also assessed the proportion of patients with CRP or ESR elevation without GCA-related signs/symptoms up to Week 26.

Results: Seventy patients were enrolled (mavrilimumab, N=42; placebo, N=28). The association of CRP or ESR elevation with unequivocal GCA-related signs/symptoms post-randomization was consistent regardless of treatment arm: 8/8 in the mavrilimumab group and 13/13 in the placebo group (Table 1). During relapse, median (range) CRP was 1.8 (1.4 – 8.4) mg/dL (mavrilimumab group) and 1.8 (1.1 – 9.0) mg/dL (placebo group). Corresponding ESR values were 39.5 (30 – 102) mm/hr (mavrilimumab group) and 49 (31 – 101) mm/hr (placebo group). Four mavrilimumab recipients had self-limited, equivocal GCA-related signs/symptoms without concurrent CRP or ESR elevation; all 4 completed the prespecified GC taper by Week 26 without need for rescue GCs, so relapse was not confirmed. At least 1 elevated CRP or ESR value in the absence of GCA-related signs/symptoms was observed in 58.8% of mavrilimumab recipients and 93.3% of placebo recipients by Week 26.

Conclusion: The observed association of CRP or ESR elevation with GCA-related signs/symptoms is consistent with the upstream mechanism and supports the utility of the stringent protocol definition of relapse. The frequency and magnitude of CRP and ESR elevations at relapse were similar in both treatment groups, suggesting that CRP and ESR remain useful in assessments of disease activity in mavrilimumab-treated patients. CRP and ESR elevations without GCA-related signs/symptoms occurred more often in placebo recipients.

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Table 1. CRP and ESR levels in patients with or without GCA relapse

Assessment [§]	Mavrilimumab N=42	Placebo N=28	Mavrilimumab N=42	Placebo N=28
	With Relapse		Without Relapse	
# of patients	8 (19.1)	13 (46.4)	34 (81.0)	15 (53.6)
Elevated CRP* or ESR†	8 (100.0)	13 (100.0)	20 (58.8)	14 (93.3)
Elevated CRP*	7 (87.5)	10 (76.9)	10 (29.4)	11 (73.3)
Median (range) mg/dL	1.8 (1.4 – 8.4)	1.8 (1.1 – 9.0)	2.6 (1.3 – 7.0)	2.0 (1.0 – 6.6)
Elevated ESR†	6 (75.0)	9 (69.2)	16 (47.1)	10 (66.7)
Median (range) mm/hr	39.5 (30 – 102)	49.0 (31 – 101)	41.5 (30 – 110)	53.5 (30 – 82)

[§]# (%), except where indicated otherwise. *CRP ≥ 1 mg/dL †ESR ≥ 30 mm/hr

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