

Case 1: Granulomatosis with Polyangiitis (GPA)

Begüm Güler

Şentürk, MD¹,

ORCID: 0000-0002-4806-317X

Assoc. Prof. Nilüfer

Alpay Kanitez, MD²

ORCID: 0000-0003-1185-5816

Birmingham Vasculitis Activity Score (BVAS) was developed to measure disease activity in patients with systemic vasculitis. The second and third versions of BVAS, which were used for the first time in 1994, were developed in 1997 and 2008, respectively [1]. On the other hand, BVAS / WG form was used for the first time in 2001, which was adapted to patients with granuloma polyangiitis (GPA) [2]. GPA is a granulomatous necrotizing type systemic small vessel vasculitis that can involve all organs, especially the upper respiratory tract and lungs. The involvement of many organs and systems, damage and activity can be seen in the same patient, and signs of infection make it difficult to measure activity in GPA. In this article, a case in which BVAS and BVAS / WG were used in clinical activity evaluation was presented and the details of the calculation were discussed.

CASE REPORT

A 40-year-old male patient presented with joint pain, weakness, anorexia, night sweats, and necrotic lesions on the hands and feet. In his history, it was learned that he had nasal obstruction and postnasal discharge for two years, increased weakness and joint pain started three weeks ago, and bruising and necrotic lesions developed in the hands and feet for 3 days. On physical examination, symmetrical polyarthritis involving the hand and foot joints and hyperemic lesions with a tendency to necrosis on the distal ends of the fingers and toes were detected (**Figure 1**). In laboratory tests, pathologically, high acute phase (C-Reactive protein: 45 mg/L, erythrocyte sedimentation rate 56 mm/hr), microscopic hematuria, high titer anti PR3 positivity [180 U/ml (<20)] were detected. Crut and purulent discharge were observed during nasal endoscopic examination. Leukocytoclastic vasculitis was detected in the skin biopsy. Thus, the diagnosis of GPA was made and 60 mg/day IV methylprednisolone was started. On the second day of the treatment, abdominal pain and hematochesia developed. Ulcers suggesting vasculitic involvement were observed in the colon during colonoscopy (**Figure 2**). Acute abdomen developed on the day of the procedure. In emergency laparotomy, it was observed that perforation developed in more than one area in the colon. Primary surgical repair was performed. At this stage, it was calculated as BVAS 22 and BVAS / WG 6 (**Figure 3**) [3]. On the post-op 2nd day, cyclophosphamide 1 g IV pulse was administered. Oral intake was opened in a short time during his follow-up, and clinical findings regressed within days. The second finger of the left hand healed with a millimeter autoamputation. The steroid dose was gradually reduced and cyclophosphamide pulse therapy was continued.



Figure 1: Purpura with a tendency to necrosis on the dorsum of the hand in a case of polyangiitis with granuloma (GPA). Severe ischemia of the third finger.

¹Koç University
School of Medicine,
Department of Internal
Medicine, Istanbul
²Koç University
School of Medicine,
Department of Internal
Medicine, Division of
Rheumatology, Istanbul



Figure 2: Broad-based mucosal ulcer seen on colonoscopy in a case of polyangiitis with granuloma (GPA).

At the fifth month visit, it was learned that he had intermittent abdominal pain, nasal crusting for 2 months, arthralgia for 15 days and severe headache. It was observed that CRP and anti-PR3, which were normalized with treatment, increased (15 mg / L, 37 U / L, respectively). A frontobasal meningeal thickening was observed on cranial magnetic resonance imaging (MRI). Cerebrospinal fluid (CSF) protein was slightly elevated. These findings were evaluated as neurological involvement of GPA. Abdominal pains were attributed to adhesions that developed after laporotomy. At this stage, it was

Is this the patient's first assessment?		Yes <input type="radio"/>		No <input type="radio"/>							
None	Active disease	None	Active disease	Persistent	New/Worse	None					
1. GENERAL						7. GENERAL					
Myalgia	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>	Arthralgia/arthritis	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>	a. arthralgia/arthritis	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>
Arthralgia / arthritis	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>	Fever (≥ 38.0 C)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	b. fever (≥ 38.0 C)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Fever ≥38° C	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>	Weight loss ≥2 kg	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>	8. CUTANEOUS	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>
Weight loss ≥2 kg	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>	a. purpura	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>
2. Cutaneous						9. MUCOUS MEMBRANES/EYES					
Infarct	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Cardiomyopathy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	a. mouth ulcers	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Purpura	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>	Congestive cardiac failure	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	b. conjunctivitis/episcleritis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Ulcer	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	c. retro-orbital mass/proptosis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Gangrene	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	d. uveitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Other skin vasculitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	e. * scleritis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
3. Mucous membranes / eyes						10. EARS, NOSE & THROAT					
Mouth ulcers	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Hypertension	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	a. bloody nasal discharge/	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>
Genital ulcers	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Proteinuria ≥1+	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	nasal crusting/ulcer	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Adnexal inflammation	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Haematuria ≥10 RBCs/hpf	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	b. sinus involvement	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Significant proptosis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Serum creatinine 125-249 µmol/L*	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	c. swollen salivary gland	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Scleritis / Episcleritis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Serum creatinine 250-499 µmol/L*	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	d. subglottic inflammation	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Conjunctivitis / Blepharitis / Keratitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Serum creatinine ≥500 µmol/L*	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	e. conductive deafness	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Blurred vision	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Rise in serum creatinine >30% or fall in creatinine clearance >25%*	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	f. * sensorineural deafness	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Sudden visual loss	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	*Can only be scored on the first assessment	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Uveitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Retinal changes (vasculitis / thrombosis / exudate / haemorrhage)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
4. ENT						11. CARDIOVASCULAR					
Bloody nasal discharge / crusts / ulcers / granulomata	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>	Headache	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	a. pericarditis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Paranasal sinus involvement	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Meningitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Subglottic stenosis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Organic confusion	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Conductive hearing loss	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Seizures (not hypertensive)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Sensorineural hearing loss	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Cerebrovascular accident	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
5. Chest						12. GASTROINTESTINAL					
Wheeze	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Spinal cord lesion	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	a. * mesenteric ischemia	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Nodules or cavities	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Cranial nerve palsy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Pleural effusion / pleurisy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Sensory peripheral neuropathy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Infiltrate	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Mononeuritis multiplex	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Endobronchial involvement	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Massive haemoptysis / alveolar haemorrhage	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Respiratory failure	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
6. Mucous membranes / eyes						13. PULMONARY					
Mouth ulcers	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	a. pleurisy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Genital ulcers	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	b. nodules or cavities	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Adnexal inflammation	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	c. other infiltrate secondary to WG	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Significant proptosis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	d. endobronchial involvement	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Scleritis / Episcleritis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	e. * alveolar haemorrhage	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Conjunctivitis / Blepharitis / Keratitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	f. * respiratory failure	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Blurred vision	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Sudden visual loss	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Uveitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Retinal changes (vasculitis / thrombosis / exudate / haemorrhage)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
7. Cardiovascular						14. RENAL					
Loss of pulses	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	a. haematuria (≥2 rbc casts (2 1+ or ≥ 10 rbc/hpf))	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Valvular heart disease	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	b. * RBCcasts	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Pericarditis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	c. * rise in creatinine > 30% or fall in creatinine clearance >25%	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Ischaemic cardiac pain	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Note: If both haematuria and RBC casts are present, score only the RBC casts (the major item)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Cardiomyopathy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Congestive cardiac failure	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
8. Renal						15. NERVOUS SYSTEM					
Hypertension	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	a. * meningitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Proteinuria ≥1+	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	b. * cord lesion	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Haematuria ≥10 RBCs/hpf	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	c. * stroke	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Serum creatinine 125-249 µmol/L*	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	d. * cranial nerve palsy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Serum creatinine 250-499 µmol/L*	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	e. * sensory peripheral neuropathy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Serum creatinine ≥500 µmol/L*	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	f. * motor mononeuritis multiplex	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Rise in serum creatinine >30% or fall in creatinine clearance >25%*	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
*Can only be scored on the first assessment	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
9. Nervous system						16. OTHER:					
Headache	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	(describe all items and * items deemed major)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Meningitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Organic confusion	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Seizures (not hypertensive)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Cerebrovascular accident	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Spinal cord lesion	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Cranial nerve palsy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Sensory peripheral neuropathy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Mononeuritis multiplex	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
10. Other						17. TOTAL NUMBER OF ITEMS:					
a.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
b.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
c.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
d.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
PERSISTENT DISEASE ONLY:						18. CURRENT DISEASE STATUS (check all that apply):					
(Tick here if all the abnormalities are due to persistent disease)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	a. Severe flare/new disease	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
References:						19. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)					
Version 1: Luggani, RA, et al. (1994). "Birmingham Vasculitis Activity Score (BVAS) in systemic necrotizing vasculitis." QJM 87(11): 671-8.						Mark line to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:					
Version 2: Luggani, RA, et al. (1997). "Disease assessment and management of the vasculitides." Baillieres Clin Rheumatol 11(2): 423-46.						Remission 0 _____ Maximum activity 10					
Version 3: Mukhyar C, et al (2008). "Modification and validation of the Birmingham Vasculitis Activity Score (version 3). Ann Rheum Dis. 2008 Dec 3. [Epub ahead of print]						20. Value in item #19: _____ (distance from 0 to tick mark in millimeters)					

BVAS: 22

BVAS-WG: 6

Figure 3: BVAS and BVAS / WG records calculated before remission induction treatment of our case. BVAS = 22 points with 1 point for arthritis, 2 points for weight loss, 6 points for skin involvement with the maximum limit, 4 points for nasal involvement, 9 points for bloody diarrhea. BVAS / WG = 6 points with 1 point each for arthritis, purpura and nasal involvement, 1x3 = 3 points for gangrene.

Is this the patient's first assessment? Yes <input checked="" type="radio"/>		No <input type="radio"/>	
None	Active disease	None	Active disease
1. General			
Myalgia	<input type="radio"/>		<input type="radio"/>
Arthralgia / arthritis	<input checked="" type="radio"/>		<input type="radio"/>
Fever ≥38° C	<input type="radio"/>		<input type="radio"/>
Weight loss ≥2 kg	<input type="radio"/>		<input type="radio"/>
2. Cutaneous			
Infarct	<input type="radio"/>		<input type="radio"/>
Purpura	<input type="radio"/>		<input type="radio"/>
Ulcer	<input type="radio"/>		<input type="radio"/>
Gangrene	<input type="radio"/>		<input type="radio"/>
Other skin vasculitis	<input type="radio"/>		<input type="radio"/>
3. Mucous membranes / eyes			
Mouth ulcers	<input type="radio"/>		<input type="radio"/>
Genital ulcers	<input type="radio"/>		<input type="radio"/>
Adnexal inflammation	<input type="radio"/>		<input type="radio"/>
Significant proptosis	<input type="radio"/>		<input type="radio"/>
Scleritis / Episcleritis	<input type="radio"/>		<input type="radio"/>
Conjunctivitis / Blepharitis / Keratitis	<input type="radio"/>		<input type="radio"/>
Blurred vision	<input type="radio"/>		<input type="radio"/>
Sudden visual loss	<input type="radio"/>		<input type="radio"/>
Uveitis	<input type="radio"/>		<input type="radio"/>
Retinal changes (vasculitis / thrombosis / exudate / haemorrhage)	<input type="radio"/>		<input type="radio"/>
4. ENT			
Bloody nasal discharge / crusts / ulcers / granulomata	<input checked="" type="radio"/>		<input type="radio"/>
Paranasal sinus involvement	<input type="radio"/>		<input type="radio"/>
Subglottic stenosis	<input type="radio"/>		<input type="radio"/>
Conductive hearing loss	<input type="radio"/>		<input type="radio"/>
Sensorineural hearing loss	<input type="radio"/>		<input type="radio"/>
5. Chest			
Wheeze	<input type="radio"/>		<input type="radio"/>
Nodules or cavities	<input type="radio"/>		<input type="radio"/>
Pleural effusion / pleurisy	<input type="radio"/>		<input type="radio"/>
Infiltrate	<input type="radio"/>		<input type="radio"/>
Endobronchial involvement	<input type="radio"/>		<input type="radio"/>
Massive haemoptysis / alveolar haemorrhage	<input type="radio"/>		<input type="radio"/>
Respiratory failure	<input type="radio"/>		<input type="radio"/>
6. Cardiovascular			
Loss of pulses	<input type="radio"/>		<input type="radio"/>
Valvular heart disease	<input type="radio"/>		<input type="radio"/>
Pericarditis	<input type="radio"/>		<input type="radio"/>
Ischaemic cardiac pain	<input type="radio"/>		<input type="radio"/>
Cardiomyopathy	<input type="radio"/>		<input type="radio"/>
Congestive cardiac failure	<input type="radio"/>		<input type="radio"/>
7. Abdominal			
Peritonitis	<input type="radio"/>		<input type="radio"/>
Bloody diarrhoea	<input type="radio"/>		<input type="radio"/>
Ischaemic abdominal pain	<input type="radio"/>		<input type="radio"/>
8. Renal			
Hypertension	<input type="radio"/>		<input type="radio"/>
Proteinuria >1+	<input type="radio"/>		<input type="radio"/>
Haematuria ≥10 RBCs/hpf	<input type="radio"/>		<input type="radio"/>
Serum creatinine 125-249 µmol/L*	<input type="radio"/>		<input type="radio"/>
Serum creatinine 250-499 µmol/L*	<input type="radio"/>		<input type="radio"/>
Serum creatinine ≥500 µmol/L*	<input type="radio"/>		<input type="radio"/>
Rise in serum creatinine >30% or fall in creatinine clearance >25%	<input type="radio"/>		<input type="radio"/>
*Can only be scored on the first assessment			
9. Nervous system			
Headache	<input type="radio"/>		<input type="radio"/>
Meningitis	<input checked="" type="radio"/>		<input checked="" type="radio"/>
Organic confusion	<input type="radio"/>		<input type="radio"/>
Seizures (not hypertensive)	<input type="radio"/>		<input type="radio"/>
Cerebrovascular accident	<input type="radio"/>		<input type="radio"/>
Spinal cord lesion	<input type="radio"/>		<input type="radio"/>
Cranial nerve palsy	<input type="radio"/>		<input type="radio"/>
Sensory peripheral neuropathy	<input type="radio"/>		<input type="radio"/>
Mononeuritis multiplex	<input type="radio"/>		<input type="radio"/>
10. Other			
a.	<input type="radio"/>		<input type="radio"/>
b.	<input type="radio"/>		<input type="radio"/>
c.	<input type="radio"/>		<input type="radio"/>
d.	<input type="radio"/>		<input type="radio"/>
PERSISTENT DISEASE ONLY: (Tick here if all the abnormalities are due to persistent disease)			
		<input type="checkbox"/>	
References:			
Version 1: Lutzman, RA, et al. (1994). "Birmingham Vasculitis Activity Score (BVAS) in systemic necrotizing vasculitis." QJM 87(11):671-8.			
Version 2: Lutzman, RA, et al. (1997). "Disease assessment and management of the vasculitides." Baillieres Clin Rheumatol 11(2): 423-46.			
Version 3: Mukhtyar C, et al (2008). "Modification and validation of the Birmingham Vasculitis Activity Score (version 3) Ann Rheum Dis. 2008 Dec 3. [Epub ahead of print]			

	Persistent	New/Worse	None
7. GENERAL			
a. arthralgia/arthritis	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>
b. fever (≥ 38.0 C)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
8. CUTANEOUS			
a. purpura	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
b. skin ulcer	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
c. * gangrene	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
9. MUCOUS MEMBRANES/EYES			
a. mouth ulcers	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
b. conjunctivitis/episcleritis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
c. retro-orbital mass/proptosis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
d. uveitis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
e. * scleritis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
f. * retinal exudates/haemorrhage	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
10. EAR, NOSE & THROAT			
a. bloody nasal discharge/ nasal crusting/ulcer	<input checked="" type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
b. sinus involvement	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
c. swollen salivary gland	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
d. subglottic inflammation	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
e. conductive deafness	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
f. * sensorineural deafness	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
11. CARDIOVASCULAR			
a. pericarditis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
12. GASTROINTESTINAL			
a. * mesenteric ischemia	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
13. PULMONARY			
a. pleurisy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
b. nodules or cavities	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
c. other infiltrate secondary to WG	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
d. endobronchial involvement	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
e. * alveolar haemorrhage	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
f. * respiratory failure	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
DETERMINING DISEASE STATUS:			
Severe flare: ≥ 1 new/worse Major item.			
Limited flare: ≥ 1 new/worse Minor item.			
Persistent disease: Continued (but not new/worse) activity.			
Remission: No active disease, including either new/worse or persistent items.			
19. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)			
Mark line to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:			
Remission	—————	—————	Maximum activity
	0	10	
20. Value in item #19: _____ (distance from 0 to tick mark in millimeters)			
14. RENAL			
a. hematuria (≥2 rbc casts) (≥ 1+ or ≥ 10 rbc/hpf)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
b. * RBC casts	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
c. * Rise in creatinine > 30% or fall in creatinine clearance >25%			
Note: If both haematuria and RBC casts are present, score only the RBC casts (the major item)			
15. NERVOUS SYSTEM			
a. * meningitis	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>
b. * cord lesion	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
c. * stroke	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
d. * cranial nerve palsy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
e. * sensory peripheral neuropathy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
f. * motor mononeuritis multiplex	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
16. OTHER: (describe all items and * items deemed major)			
	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
17. TOTAL NUMBER OF ITEMS:			
a.	b.	c.	d.
1			
Major New/Worse	Minor Persistent	Major New/Worse	Minor Persistent
18. CURRENT DISEASE STATUS (check all that apply):			
a. Severe flare/new disease	()		
b. Limited flare/new disease	()		
c. Persistent severe disease	()		
d. Persistent limited disease	()		
e. Remission	()		

BVAS: 7

BVAS-WG: 5

Figure 4: BVAS and BVAS / WG records calculated in our case's 5th month visit. BVAS 7 points with 1 point for arthralgia, 2 points for nasal findings, 1 point for headache and 3 points for meningitis. BVAS / WG 6 points with 1 point for arthralgia, 1 point for nasal involvement, 1x3 = 3 points for meningitis.

BVAS 7 and BVAS / WG 5 (Figure 4). The methylprednisolone dose was increased to 36 mg / day and rituximab (1 g every two weeks) was started. Maintenance treatment with rituximab monotherapy continues in our patient who completed his second year in remission. Last calculated BVAS and BVAS / WG is "0".

DISCUSSION

In the BVAS form, any involvement associated with the diagnosis of vasculitis at the initial evaluation is marked as new / worsening finding. Each finding marked has a different numerical equivalent. In our case, all the involvements for the first BVAS and BVAS / WG were marked as new / worsening and the corresponding scores were collected. One of the major differences between BVAS and BVAS / WG is that nasal involvement scored 4 points in BVAS and 1 in BVAS / WG. Another difference is that the contribution of the same involvement to the score varies depending on whether there is a persistent or new / worsening finding in BVAS, whereas such a distinction does not contribute to the score in BVAS / WG. Differently, the involvements expressed as major involvement are scored with x3. Another issue to be aware of is that although each finding is scored separately, each system has a maximum value of the total score. Although the sum of skin involvement separately was 8 in the BVAS that was first calculated in our case, the result is 6 points, which is the maximum score that can be obtained. The maximum scores for each section are also available in the forms.

It is important to distinguish between persistent and new and worsening findings in the scoring we made at the fifth month visit. The adhesions formed in the intestine were evaluated as permanent damage and were not marked in BVAS and BVAS / WG. Since the presence of nasal findings exceeded 28 days, it was considered to be persistent. Thus, while the contribution of nasal involvement to the total score in BVAS decreased by 2 points, it did not change in BVAS / WG.

KEY MESSAGES

- *At GPA, evaluating activity with BVAS and BVAS / WG benefits clinical decisions as well as academic research.*
- *When calculating BVAS and BVAS / WG, it is important to correctly assess the relationship of the findings with the disease, whether damage or persistence.*
- *It should not be overlooked that BVAS and BVAS / WG scores are different according to the systems with which clinical findings are related, and there is a maximum score limit for each system in BVAS.*

References

1. Mukhtyar C, Lee R, Brown D, et al. Modification and validation of the Birmingham Vasculitis Activity Score (version 3) Ann Rheum Dis. 2009; 68:1827–1832.
2. Stone JH, Hoffman GS, Merkel PA, et al. A disease-specific activity index for Wegener's granulomatosis: modification of the Birmingham Vasculitis Activity Score. International Network for the Study of the Systemic Vasculitides (INSSYS). Arthritis Rheum. 2001; 44:912–920.
3. Bes C, Karadağ Ö. Birmingham Vaskülit Aktivite Skorları (Birmingham Vasculitis Activity Scores, BVAS and BVAS-WG). <http://www.vaskulit.hacettepe.edu.tr/belge/bvas121119.pdf>