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.

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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 laparotomy. At this stage, it was

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

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**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.

**BVAS: 22**

**BVAS-WG: 6**
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

