

Chronic large nasal bloody crusting and recurrent episcleritis: Limited granulomatosis with polyangiitis

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Granulomatosis with polyangiitis (GPA) (formerly named Wegener's granulomatosis) is an uncommon kind of systemic vasculitis involving small-to-medium sized vessels, and categorized as ANCA-associated vasculitis with the presence of anti-neutrophil cytoplasm antibodies (1). It is characterized by the formation of necrotizing granuloma in the upper and/or lower respiratory tract and glomerulonephritis. Subsequently, it affects almost any organ or tissue. GPA is influenced by genetic, immunologic, and environmental factors (2). GPA affects people at any age, usually between 60 and 70 year of age in both sexes. Two forms of GPA are systemic and diffuse forms; systemic GPA typically includes renal and pulmonary manifestations and/or vital organ involvement and systemic symptoms, such as fever, anorexia, or weight loss, and as localized/limited forms that predominantly affect the upper respiratory tract, but they are recurrent (3). The diagnosis of disease is based on the classification criteria for granulomatosis with polyangiitis provided by the American College of Rheumatology (4). Currently, ANCA is used for diagnosis in clinical practice.

A 69-year-old female with painless, redness in the left eye, which persisted for 2 weeks, and chronic weakness and general malaise, was referred to the rheumatology clinic for further evaluation of her condition. She had ophthalmic history, including an episcleritis requiring systemic corticosteroid treatment 2 years ago. Medical history revealed that she has been suffering from chronic nasal dryness and nasal bloody crusting rhinore/discharge (Figure 1) since 5 years. Her ophthalmic and ear nose, and throat (ENT) examination after admission showed episcleritis and nasal purulent crusting. There was no evidence of other organ/tissue involvement.



Her laboratory findings revealed systemic inflammation. Blood parameters were as follows: white blood cell count $11 \times 10^3/\mu\text{L}$, neutrophil 81%, C-reactive protein 84 mg/dl, erythrocyte sedimentation rate 89 mm/h. Urine analysis result was normal. Anti-PR3, anti-MPO, and antinuclear antibodies were negative. Rheumatoid factor was 75.6 IU (normal value: <20 IU).

Sinusal tomography showed increased amount of soft tissue in the paranasal sinuses. Chest graphy and computed tomography results were normal. Biopsy of the nasal mucosa and the histopathological findings were consistent with GPA (Figure 2-4).

Nasal-sinus involvement occurs in approximately 85% of patients with GPA, such as bloody nasal discharge or crusts, chronic sinusitis, bone, and/or cartilage destruction (5, 6). The ocular manifestations, such as conjunctivitis, episcleritis, keratitis, scleritis, uveitis, and retinal vasculitis, can occur in approximately half of the patients. Here, the patient presented with chronic nasal symptoms and recurrent episcleritis, and histopathological granulomatous manifestations of nasal mucosa without evidence of lung and kidney disease, and demonstrated the presence of limited form GPA with absence of ANCA. Histopathological examination is essential for diagnosing, particularly in ANCA-negative patients; how-



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Figure 1. Large nasal discharge with bloody purulent appearance

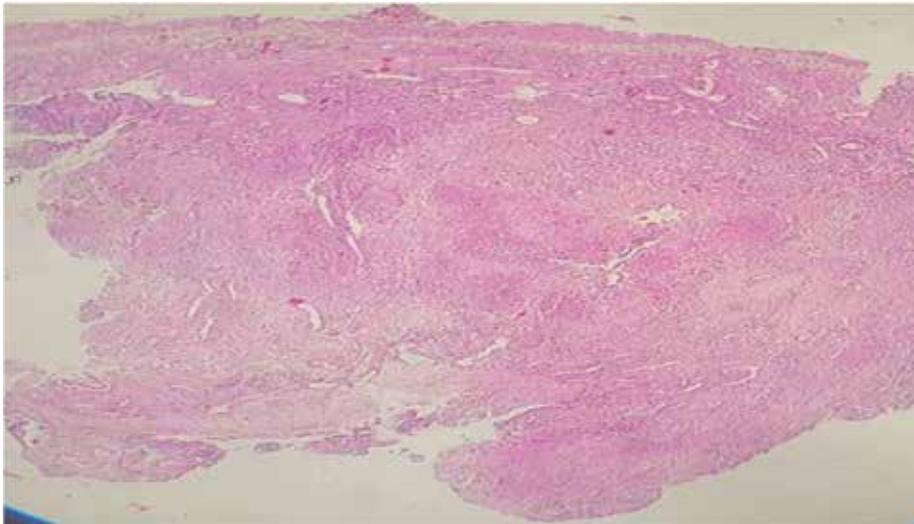


Figure 2. Scanning magnification of biopsy material. Respiratory mucosa is present on upper part of biopsy. A prominent inflammatory infiltration is seen on full thickness of biopsy material (X2, Hematoxylen&Eosin).

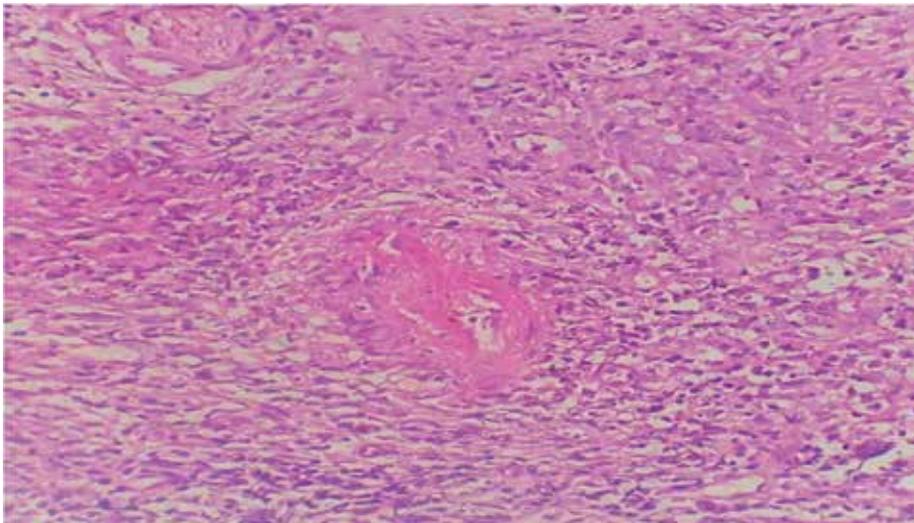


Figure 3. Medium magnification of biopsy material shows a section of vascular structure (X20, Hematoxylen&Eosin). Fibrinoid necrosis is present on intimal part of vessel.

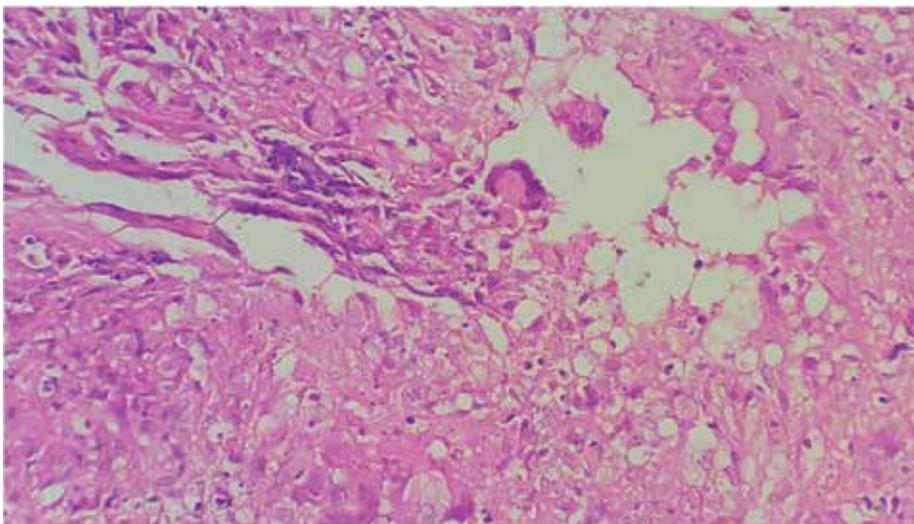


Figure 4. Higher magnification of inflammatory infiltration shows multinucleated giant cells (40X, Hematoxylen&Eosin).

ever, the absence of ANCA does not exclude this diagnosis. ANCA may not be present in cases of limited form of GPA (7). Current treatment comprised corticosteroid and cyclophosphamide until disease remission, followed by a less toxic immunosuppressant, such as azathioprine (8).

Clinical presentation of primary vasculitis can be variable; therefore, careful attention needs to be paid to patient's anamnesis, clinical examination, and laboratory findings. Our aim was to show the importance of nasal bloody crusting in the diagnosis of vasculitis.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

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