Clinical Case Study

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A CASE OF WEGENER'S GRANULOMATOSIS PRESENTING WITH GASTROINTESTINAL BLEEDING

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Wegener's granulomatosis (WG) was first described by Wegener in 1936 (Srouji, Andrews, Edwards, & Lund, 2006). As an autoimmune systemic disease, it is the idiopathic systemic form of vasculitis characterized by the presence of necrotizing granulomas and vasculitis in the upper airways, lower airways, and kidneys. This disease can affect any organ system (Lyons et al., 2012). This case presents a patient with WG presenting with gastrointestinal (GI) bleeding that is rarely reported in medical literature.

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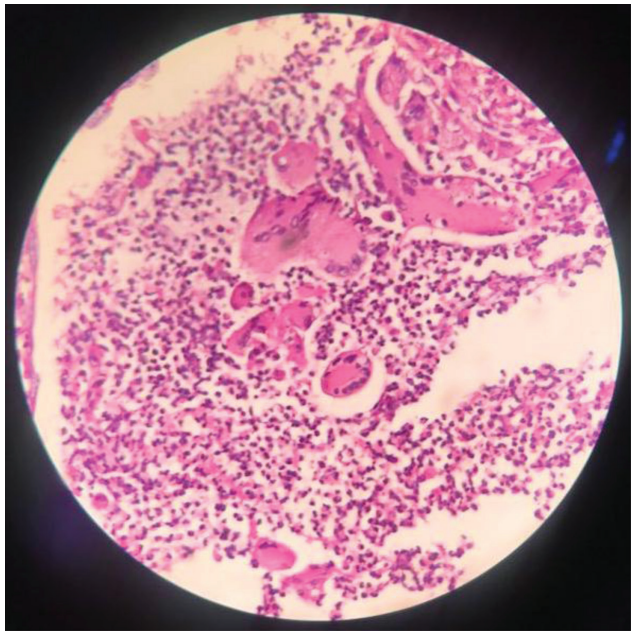

FIGURE 1. Histopathology of nasopharyngeal mucosa showing granuloma.

Case Report

A 23-year-old young man was diagnosed with WG 2 years ago. At that time, he presented with cough, nasal congestion, and joint pain. His chest computed tomographic (CT) scan showed that the right and left lobes of the lung had multiple soft-tissue masses. He was diagnosed with pulmonary infection, admitted, and treated with antibiotics for 2 weeks. The treatment was not effective. A nasopharyngeal CT scan was then obtained. It showed that the nasopharynx was normal. Nasopharyngoscopy showed nasopharyngeal mucosal ulceration, and histopathology showed granuloma (Figure 1). Erythrocyte sedimentation rate was 51 mm/hour. C-reactive protein was 47 mg/L. Routine microscopy of urine revealed 3+ proteinuria and red blood cell (RBC) casts. Antinuclear antibody, rheumatoid factor, and T-SPOT were negative. However, cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA) and proteinase-3 were positive. Viral markers for HIV infection, hepatitis B and hepatitis C, and syphilis were nonreactive. The remaining routine hematological and biochemical investigations were normal. On the basis of symptoms, laboratory examinations, CT scan, and histopathology, as well as the diagnostic criteria of WG, the patient was diagnosed with WG.

Intravenous methylprednisolone (160 mg/day) and cyclophosphamide (400 mg once weekly) were administered, and the clinical symptoms of cough and nasal congestion alleviated within a week, followed by the reduction in joint pain in 2 weeks. The patient was treated with low-dose corticosteroids and mycophenolate mofetil to maintain remission. Urinalysis was normal after 4 weeks.

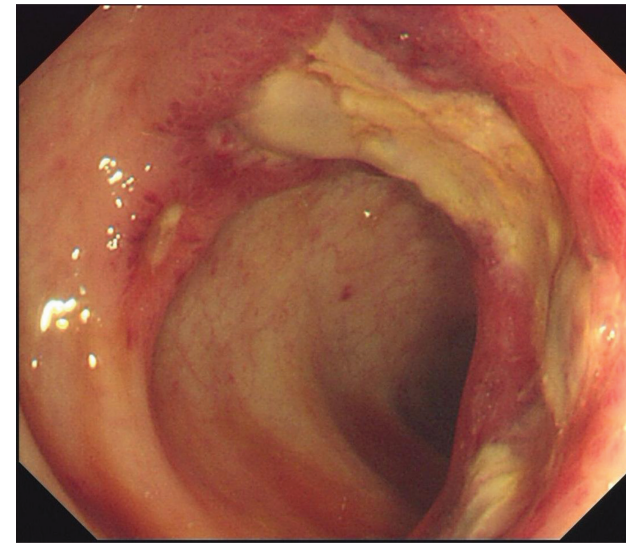

FIGURE 2. Findings of colonoscopy before the treatment (abnormal mucosal pattern, erosions, and ulcerations).

The patient had recurrent intermittent abdominal pain and melena 2 years after the initiation of immunosuppressive treatment. On Day 5 after admission, he experienced hematochezia. His hemoglobin level was normal. The chest CT scan showed reduction in soft-tissue masses. Gastroscopy was normal. Colonoscopy was performed, and erosions and ulcerations were found in the terminal ileum and colon (Figure 2). Histological analyses demonstrated chronic mucosal inflammation. Combined with detailed medical history, laboratory examination, abdominal CT, and endoscopy, we assumed his GI bleeding was related to WG. So, we used methylprednisolone for this patient; hematochezia stopped after 3 days. The patient was prescribed prednisone at time of discharge from the

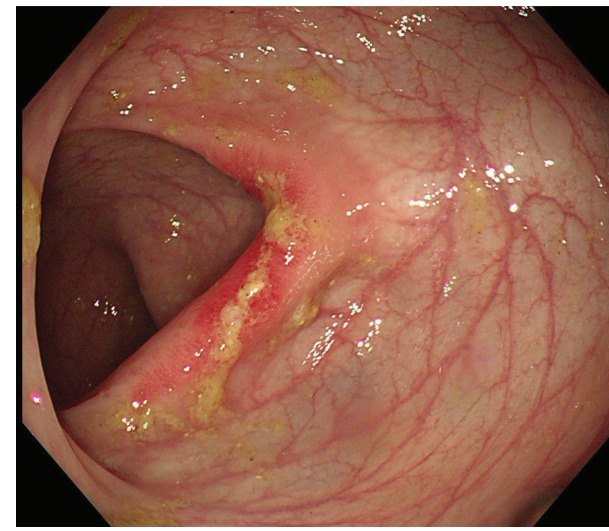

FIGURE 3. Findings of colonoscopy after treatment (the ulcer is much smaller).

hospital. Colonoscopy was repeated 3 months later. The colonic ulcers were smaller or had healed after treatment (Figure 3). Pathology still showed chronic mucosal inflammation; therefore, treatment of WG was continued.

Discussion

Wegener's granulomatosis is an uncommon autoimmune systemic disease that affects different body systems. Antibodies against cytoplasmic neutrophilic granules (c-ANCA) are present in 80% of patients (Kallenberg, Leontine Mulder, & Cohen Tervaert, 1992). Among the various symptoms, the upper airways, lung, and kidneys are the most frequent manifestations of WG. However, it rarely involves organs of the GI system and usually occurs long after the onset of initial symptoms. Gastrointestinal involvement has been described in 10%–24% of patients with WG (Izzedine, Lacaille, & Deray, 2001), with the most frequent GI symptoms being abdominal pain, bleeding, and diarrhea (Cabral et al., 2009). In this case, the patient met the American College of Rheumatology 1990 criteria for the classification of WG and had a positive test for c-ANCA (Leavitt et al., 1990).

The case is consistent with the review of literature in that bleeding of the GI tract occurred 2 years after the initial diagnosis had been established. A close follow-up is required to monitor the relapse and determine the involvement of other organs.

Acute GI bleeding can be an emergency with a high mortality rate and can be lethal if left untreated. Early diagnosis and treatment are very important. When WG involves the GI tract or there is bleeding, we often do not consider WG as the differential diagnosis. Inflammatory bowel disease, lymphoma, and other colonic disease need to be considered. Many times, the diagnosis is dependent on histopathology. Although it may be difficult for the pathologist, we need detailed clinical information and multiple biopsies to make the diagnosis.

In this case, the patient presented with hematochezia. We considered multiple differential diagnoses of lower GI bleeding including inflammatory bowel disease, tuberculosis, tumor, and hemorrhoids. The patient was taking immunosuppressants for a prolonged time period, but we needed to eliminate the diagnosis of tuberculosis and tumor. Colonoscopy revealed diffuse colonic involvement with erosions and ulcers. Biopsies from the ulcer showed nonspecific ulceration without malignancies, colonic crypts, or granuloma formation. Urine showed a large amount of RBCs, and a positive test for c-ANCA. These laboratory results determined that WG was active. Hematochezia was associated with WG vasculitis.

From the reports in the literature, remission can be achieved in up to 90% of cases with the help of therapeutic agents such as steroids and other immunosuppressant drugs such as cyclophosphamide (Kuwahara et al., 2006). This patient was effectively treated with corticosteroids.

Conclusion

Wegener's granulomatosis is a systemic vasculitis and can involve multiple organs. When WG involves the GI tract, it can lead to abdominal pain, melena, even shock bleeding. Along with examination, endoscopy and biopsies should be carried out for patients with suspected WG. Corticosteroid is an effective treatment option for most patients, but surgery for the bleeding or bowel perforation may be needed.

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