

Microscopic Polyangiitis Disguised in Gastrointestinal Bleed: A Rare Association

Abstract:

Microscopic polyangiitis (MPA) is an idiopathic autoimmune disease characterized by necrotizing vasculitis without granulomatous inflammation that predominantly affects small vessels. It most commonly presents in elderly patients but can occur at any age. Here we present a case of an elderly patient who presented with exertional dyspnea and fatigue in the setting of anemia and was diagnosed with microscopic polyangiitis. The patient had a past medical history significant for chronic kidney disease stage IIIb with worsening creatinine over the last four months along with feeling fatigued and intermittently dyspneic associated with black stools for the previous three weeks. A kidney biopsy revealed fibro-cellular crescents consistent with microscopic polyangiitis, and the patient was started on intravenous pulse dose steroids and later put on hemodialysis for worsening creatinine. He was later discharged home on rituximab and oral steroids. MPA is a rare vasculitis that presents with renal dysfunction and occasionally with pulmonary involvement. Gastrointestinal bleeding is rarely associated with the MPO and is thought to be caused by arterial aneurysms. Therefore, MPO should be considered in mind while evaluating GI bleeding in patients with worsening kidney function.

Introduction:

Microscopic polyangiitis (MPA) is an idiopathic autoimmune disease characterized by necrotizing vasculitis without granulomatous inflammation that predominantly affects small vessels. (1) It most commonly presents in elderly patients in their 50-60s but can occur at any age and is rare in children with equal distribution in both male and female patients. (2) MPA is predominantly observed in Asian countries, particularly in China and Japan (3), but studies in the United States showed a higher incidence (at least two-fold) in the white population as compared to other ethnicities. (4) The commonly affected organ systems are respiratory and renal, but small blood vessels in any organ or tissue can be involved. (5) Gastrointestinal system involvement is quite rare, and here we present a case of an elderly patient who presented with anemia and gastrointestinal bleeding and was diagnosed with microscopic polyangiitis.

Case Presentation :

Our patient is an 83-year-old male with a past medical history significant for hypertension, hyperlipidemia, and chronic kidney disease stage III-b was referred to the hospital by his primary care physician when he was found to be having abnormal lab results concerning anemia and progressively worsening creatinine over the last 4 months. He reported feeling fatigued and dyspneic on exertion associated with loose black stools for the last 2-3 weeks. The patient didn't have any prior history of colonoscopy or esophagogastroduodenoscopy (EGD). On admission, he was hemodynamically stable with the labs remarkable for hemoglobin of 7g/dL and serum creatinine of 4.92 (baseline 1.6). Fecal occult blood testing (FOBT) was positive, and the urinalysis was concerning for micro-hematuria without

any signs of infection. The patient was transfused a unit of blood and was started on intravenous pantoprazole. An extensive workup for intrinsic renal disease was sent, including free light chain, anti-neutrophilic cytoplasmic antibodies, hepatitis panel, rheumatoid factor, and complement levels. EGD was done that revealed non-bleeding erosive gastropathy and non-bleeding duodenal ulcer with a clean ulcer base. The p-ANCA level was elevated concerning for underlying vasculitis or autoimmune etiology. The kidney biopsy revealed fibro-cellular crescents consistent with microscopic polyangiitis. (figure 1) The patient was started on intravenous pulse dose steroids and was put on hemodialysis over the next few days for worsening renal function. He was switched to rituximab and oral steroids and was able to come off dialysis in the next few days, and was discharged on oral steroids with outpatient nephrology follow-up.

Date	WBC	Hemoglobin	Platelets	Creatinine
On Admission	10.6	7	142	4.92
Day 1	9.4	8.1	123	5.22
Day 3	11.7	7.3	160	6.05
Day 7	16	7.9	187	4.15
Day 10	13	8.3	209	3.88
On Discharge	9.0	7.7	261	2.70

Table 1: Labs during hospitalization stay

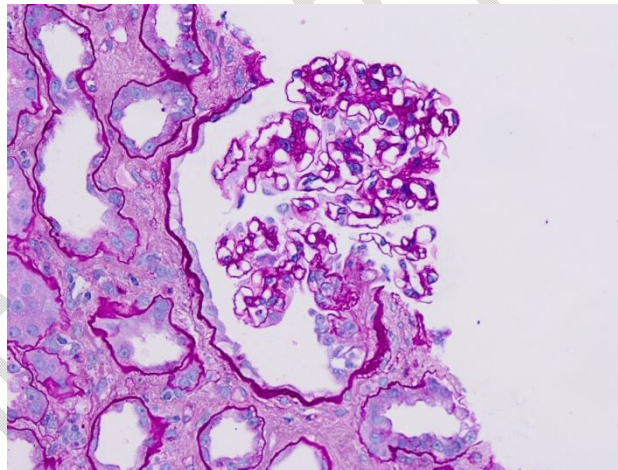


Figure 1: Fibro-cellular crescents consistent with Microscopic Polyangiitis

Discussion:

MPA is a rare vasculitis that presents most commonly with renal dysfunction and occasionally with pulmonary involvement. Although gastrointestinal bleed is frequently associated with medium vessel vasculitis, rare cases associated with the MPO have also been identified. (6) A literature review showed that gastrointestinal manifestations can be present as soon as three months after the diagnosis and are thought to be caused by arterial aneurysms. Therefore, MPA should be kept in mind while evaluating GI bleeding in patients with worsening kidney function. The diagnosis must be established by testing for MPO-ANCA with ELISA due to higher specificity. (7) Following the laboratory testing, the diagnosis should be confirmed with a biopsy from the active disease site.

The therapeutic goal is to achieve a long-standing remission and consists of an initial induction phase putting the active disease into remission followed by a remission phase to prevent relapse. Immunosuppressive therapy is recommended in all patients with active disease. (8) The recommended induction therapy consists of glucocorticoids in combination with rituximab or cyclophosphamide, with rituximab preferred over cyclophosphamide given its better efficacy and safety profile. (9) The same approach was adopted in our patient where he was started on pulse dose steroids and was later switched to rituximab. Following induction therapy, remission is maintained with rituximab, azathioprine, methotrexate, or mycophenolate.

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