Multiorgan Involvement with Severe Gastrointestinal System Manifestations in a Patient with Granulomatosis with Polyangiitis

Granulomatosis with polyangiitis (GPA) is a systemic vasculitis affecting small to medium-sized blood vessels of various organ systems with necrotizing and granulomatous inflammation. It most commonly involves the respiratory tract and kidneys, and involvement of other organ systems is less common. When kidneys are affected, it has high predilection to cause end-stage renal disease (ESRD). Gastrointestinal (GI) involvement happens rarely in GPA but when affected, there is high risk of morbidity and mortality from intestinal perforation and bleeding.

In the current issue of the journal, Bagai *et al*. describe a case of 50-year-old male with GPA involving multiple organ systems and severe GI manifestations.[1] He initially presented with ear-nose and throat (ENT) manifestations but progressed to have diffuse multisystemic involvement affecting the skin, lungs, GI system, and kidneys. The diagnosis was made by skin biopsy, surgical pathology of the resected bowel, and a positive c-ANCA (antineutrophil cytoplasmic antibody) titer. GI manifestations included severe lower GI bleeding, development of terminal ileal stricture and ulceration, multiple colonic ulcers, and multiple small bowel telangiectasias. He was treated with intravenous (IV) steroids, cyclophosphamide, hemodialysis, and plasmapheresis. Surgical resection was also attempted but the patient eventually died.

GI involvement has been described in most systemic small vessel vasculitides such as GPA, microscopic polyangiitis, eosinophilic granulomatous with polyangiitis, Henoch Schonlein purpura, and cryoglobulinemic vasculitis, although with variable frequencies.[2] GI manifestations are often indistinguishable from those of inflammatory bowel disease, and the patient may present with GI bleeding, ulceration or perforation. Often GI symptoms are the first manifestations that predate involvement of other organ systems and directly correlate with the adverse patient outcomes.[3]

The authors highlight the catastrophic GI involvement in ANCA vasculitis. Although their patient's clinical and laboratory features fit the diagnostic criteria for GPA,[4] other multi-system small vessel vasculitides can have a very similar presentation. Patients with GPA commonly present with ENT or respiratory manifestations (up to 90%).[5] Renal involvement is seen in 18% at the onset and in over 85% eventually happens during the course of the illness.[6] In a recent study by Sharma *et al*., GI involvement in GPA was seen in 12.3% of patients[7] and a presence of GI or renal involvement was noted to predict a worse outcome in the multivariate analysis. In another study,[8] GI involvement was noted in 9 of the 34 (26%) patients with GPA during the course of illness. GI bleeding or perforation was seen in six patients and two required surgical intervention.

Clinical features in conjunction with laboratory parameters are very important in making a specific diagnosis. Patient in the current report had severe acute kidney injury, active urine sediment along with GI bleeding and perforation. Pathology of the resected bowel showed necrotizing inflammation, commonly seen in other vasculitides. Skin biopsy confirmed the presence of small vessel vasculitis and a positive c-ANCA clinched the diagnosis of GPA. The patient was treated with IV steroids, cyclophosphamide, and plasmapheresis keeping in line with the current recommendations for organ-threatening ANCA vasculitis.[9] An exploratory laparotomy led to surgical resection of the involved small bowel. Despite potent immunosuppressive medications and extensive surgical resection, the patient could not be saved and succumbed to the complications of renal failure, GI bleeding, and perforation.

In conclusion, GI involvement in GPA is uncommon but not unknown and can have a catastrophic presentation. High clinical suspicion with early aggressive immunosuppressive therapy and timely surgical intervention remains the cornerstone of management.

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