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Case report

Microscopic polyangiitis: Atypical presentation with extensive small bowel necrosis, diffuse alveolar hemorrhage, and renal failure

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ABSTRACT

Microscopic polyangiitis is an uncommon systemic vasculitis of varying severity that is associated with myeloperoxidase (MPO) and perinuclear antineutrophil cytoplasmic (p-ANCA) antibodies. The most commonly affected organs are the lungs and kidneys. We report on a very unusual case of microscopic polyangiitis presenting with severe mesenteric ischemia in addition to diffuse alveolar hemorrhage and acute renal failure. The patient was initially diagnosed with acute pancreatitis at an outside facility given his severe abdominal pain and elevated pancreatic enzymes. Further investigations after transfer to our facility determined that the patient was actually suffering from a severe exacerbation of previously diagnosed microscopic polyangiitis. He quickly developed diffuse alveolar hemorrhage (DAH) necessitating intubation and acute kidney injury (AKI) requiring dialysis. He subsequently developed mesenteric ischemia and bowel necrosis resulting in emergent laparotomy and extensive small bowel resection. Physicians need to be aware that microscopic polyangiitis can very rarely present with severe involvement of the abdominal viscera and mesenteric vessels. Severe disease necessitates the use of high dose IV steroids, rituximab or cyclophosphamide, and plasma exchange (PLEX).

1. Introduction

Microscopic polyangiitis is one of the pulmonary-renal vasculitides syndromes associated with MPO and p −ANCA antibodies. It typically results in pulmonary and renal involvement characterized by diffuse alveolar hemorrhage (DAH) and acute kidney injury (AKI) respectively; whereas involvement of other organ systems is rare. We report on a rare case of microscopic polyangiitis that presented initially with severe mesenteric ischemic resulting in bowel necrosis. The patient quickly developed multiorgan failure with severe pulmonary and renal dysfunction as described below.

2. Case report

A 56 year old male patient with a 2 month history of constitutional symptoms presented to an outside hospital with abrupt abdominal pain and was initially diagnosed with acute pancreatitis (Fig. 1). His respiratory status declined rapidly thereafter necessitating intubation and transfer to our facility. On admission, a chest x-ray (Fig. 2) and chest computed tomography (CT) (Fig. 3) showed diffuse bilateral pulmonary infiltrates, and an EKG revealed new onset atrial fibrillation. Bronchoalveolar lavage performed via bronchoscopy revealed a progressively bloody return indicating DAH. He developed acute kidney injury (AKI) and his serum creatinine eventually peaked at 6.5 mg/dL. Urinalysis showed proteinuria and RBC casts. Further history was subsequently obtained from family members who confirmed that the patient had been previously diagnosed with microscopic polyangiitis via a kidney biopsy and had been put on oral cyclophosphamide by his home nephrologist. However, he had been non-compliant due to financial constraints. His kidney biopsy slides were obtained for review by our pathologists and the diagnosis of ANCA associated vasculitis was reconfirmed. The results of the kidney biopsy in combination with the positive antibodies for MPO and p-ANCA

Abbreviations: MPO, Myeloperoxidase; ANCA, Anti-neutrophil cytoplasmic autoantibody; DAH, Diffuse alveolar hemorrhage; AKI, Acute kidney injury; PLEX, Plasma exchange.

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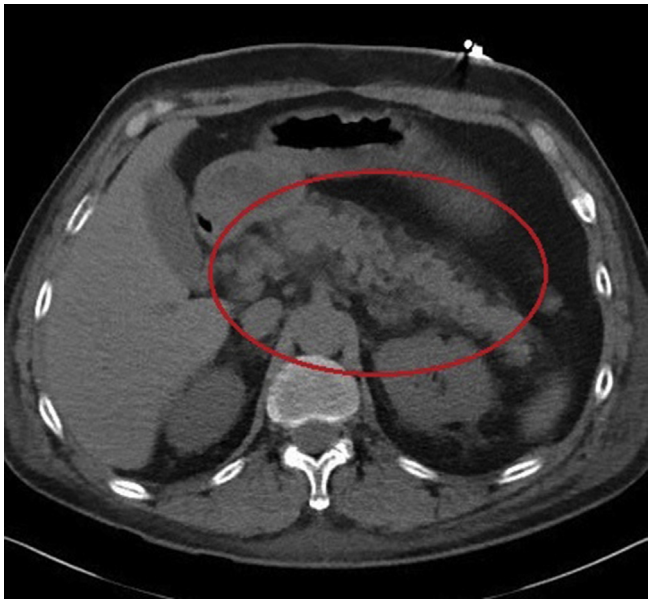

Fig. 1. CT Abdomen showing pancreatitis with diffuse peripancreatic edema.

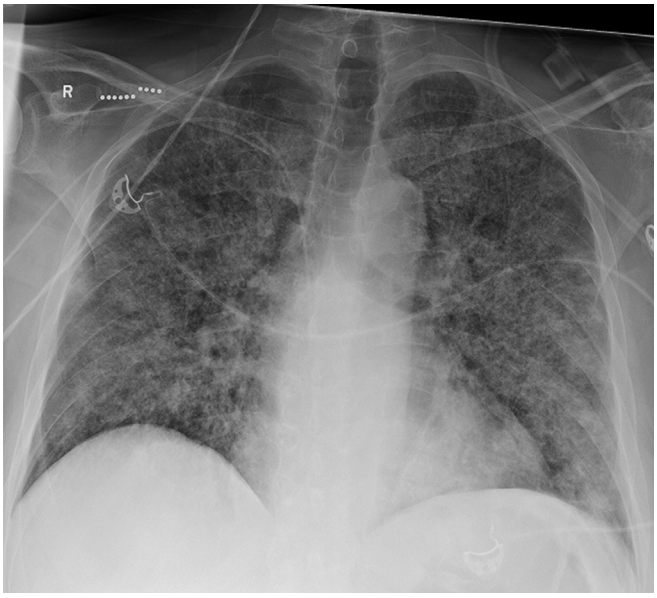

Fig. 2. Chest X-ray showing diffuse bilateral interstitial and fluffy (alveolar) type infiltrates in both lung fields.

confirmed the diagnosis of microscopic polyangiitis. High dose intravenous steroids, Rituximab, plasma exchange (PLEX), and hemodialysis were initiated with considerable and rapid improvement allowing for extubation. On day 6, however, he developed acute abdominal distention and respiratory distress leading to reintubation. Abdominal CT showed extensive small bowel ischemia (Fig. 4). An emergent laparotomy was performed and resulted in removal of 60% of the terminal small intestine, placement of an end ileostomy, and initiation of total parenteral nutrition. Mesenteric surgical pathology revealed microthrombi but no evidence of vasculitis. His abdomen was temporarily closed and he underwent subsequent end ileostomy revision and abdominal washout. He continued to improve and was ultimately discharged after a prolonged hospital stay including 2 weeks in the ICU and one

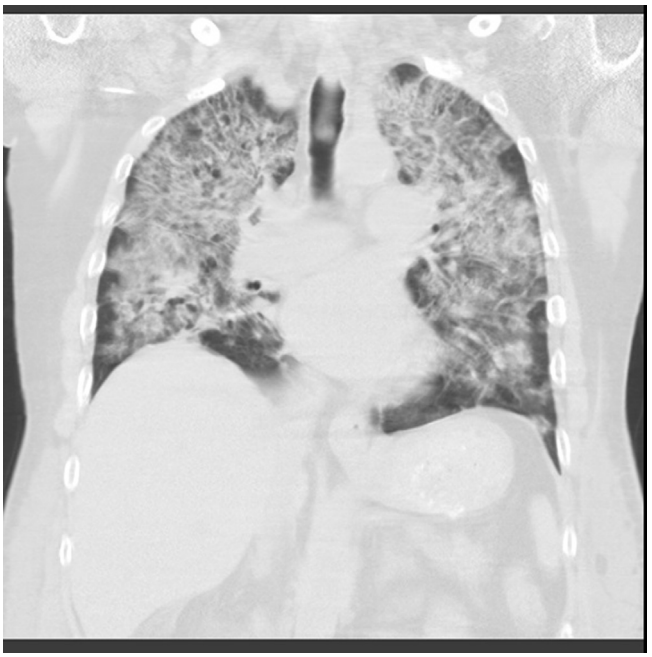

Fig. 3. Coronal Chest CT scan image with diffuse bilateral ground glass infiltrates and focal areas of consolidation.

month of dialysis. Specific microscopic polyangiitis treatment included a total of 6 days of high dose IV methylprednisolone, 7 sessions of PLEX, and 4 doses of Rituximab. This was followed by a 1 mg/kg/day oral prednisone taper with rheumatology and nephrology outpatient follow-up.

3. Discussion

Microscopic polyangiitis is a pauci-immune, ANCA-associated systemic vasculitis that affects small blood vessels. It typically involves the lungs and kidneys, with rare potential to involve any organ. Manifestations in the lungs typically include interstitial pneumonitis or DAH. The kidneys can develop necrotizing crescentic glomerulonephritis with subsequent acute renal failure [1,2].

Microscopic polyangiitis initially presents with non-specific symptoms like fever, fatigue, arthralgia, cough, and hemoptysis, with renal failure being a late manifestation. ANCA tends to be detectable at symptom onset making it useful for early diagnosis and treatment. Persistent or intermittent ANCA detection is an independent risk factor for relapsing disease but treatment for asymptomatic disease is controversial [3].

The etiology of anti-neutrophil cytoplasmic antibodies (ANCA) is thought to be caused by antigenic mimicry from multifactorial triggers. Some of the more common associations and hypothesized triggers include infections like Coxsackie B3 and parvovirus B19, silica, and medications like PTU, hydralazine, and allopurinol [3]. ANCA targets antigens found in the cytoplasmic space of polymorphonuclear (PMN) leukocytes. C-ANCA has cytoplasmic fluorescence reactivity to proteinase 3 (PR3), whereas P-ANCA has perinuclear fluorescence reactivity to myeloperoxidase (MPO). P-ANCA and MPO are most associated with microscopic polyangiitis [3].

There are conflicting studies looking at the possible pathogenic role of ANCA, but the evidence that supports the theory of ANCA's pathogenicity include animal models showing that P-ANCA may induce vasculitis in immunodeficient mice, ANCA titers correlating