A Girl with End-Stage Kidney Disease Due to Granulomatosis with Polyangiitis Presenting with Gastrointestinal Bleeding “a rare presentation”

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Granulomatosis with polyangiitis (GPA) is an uncommon vasculitis affecting small- and medium-sized vessels, predominantly in adults (average age approximately 45 years) and infrequently in children. It can involve multiple organ systems.
In patients with end-stage kidney disease (ESKD) due to GPA, disease activity is often tends to wane after the commencement of dialysis.
We report a 12-year-old girl with ESKD due to GPA who presented with severe gastrointestinal (GI) bleeding and profound anemia, owing to diffuse microaneurysms in the small bowel (particularly the ileum) and colon—an exceedingly rare manifestation. She responded dramatically to induction therapy comprising methylprednisolone pulse, plasmapheresis, rituximab followed by maintenance therapy with prednisolone and mycophenolate mofetil.
This case highlights that while the recurrence risk of ANCA-associated vasculitis in dialysis patients is low, the possibility remains and may present with atypical extrarenal involvement. Given the elevated risk of infection with immunosuppression in ESKD patients, maintenance immunosuppressive therapy may be discontinued in the absence of extrarenal disease—but this decision necessitates careful consideration.

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Case Report

INTRODUCTION

Granulomatosis with polyangiitis (GPA) is a rare vasculitis affecting small- and medium-sized vessels that can involve individuals of any age, though it most commonly occurs in adults (mean age approximately 45 years) and less frequently in children. In adult populations, a slight male predominance is reported, whereas in pediatric cases a modest female preponderance is observed. GPA may involve nearly all organ system, with renal and pulmonary involvement being among the most common manifestations. In patients who progress to end-stage kidney disease (ESKD) resulting from GPA, disease activity is generally considered to decline after dialysis initiation. We present here an unusual presentation of GPA in a pediatric patient with ESKD who developed severe gastrointestinal (GI) bleeding due to diffuse intestinal microaneurysms—a rarely described complication.

CASE PRESENTATION

A 12-year-old girl with known ESKD attributable to GPA (disease onset at age 9 with fever, weakness, fatigue, hematuria, proteinuria and renal biopsy confirming GPA), received hemodialysis after progressing to ESKD at age 11. Her immunosuppressive regimen consisting of mycophenolate mofetil and prednisolone, was gradually tapered, while a low maintenance dose of

prednisolone continued. During routine follow-up she remained asymptomatic until her presentation to the emergency department with a 3-day history of fever, diarrhea, severe abdominal pain, anorexia and pallor. On examination: temperature 39 °C, blood pressure 100/70 mmHg, pulse 140/min, respiratory rate 30/min, oxygen saturation 89% on room air (95% with supplemental oxygen). She appeared ill, pale, with generalized abdominal tenderness, guarding and rebound tenderness. Laboratory investigations revealed leukocytosis with neutrophil predominance, erythrocyte sedimentation rate (ESR) 80 mm/h, C-reactive protein (CRP) > 200 mg/L, blood urea nitrogen (BUN) 38 mg/dL, serum creatinine (SCr) 9 mg/dL, albumin 2.8 g/dL. Electrolytes, glucose, alanine aminotransferase (ALT), aspartate aminotransferase (AST), Alkaline phosphatase (ALP), D-dimer, amylase, lipase, calcium, phosphorus, magnesium were within normal limits. Stool examination showed RBCs and WBCs ~40–50 per high-power field. Abdominal/pelvic ultrasound revealed several non-specific lymph nodes in the right lower quadrant. Surgical consultation and contrast CT scan of the abdomen demonstrated marked thickening of the transverse and descending colon consistent with infectious colitis vs vascular colitis. Broad-spectrum intravenous antibiotics (meropenem, ciprofloxacin, metronidazole) were started for presumed infectious colitis. Because of persistent pain, upper endoscopy and colonoscopy were performed, which revealed multiple erosions in the antrum and aphthous lesions throughout the colon. Cytomegalovirus (CMV) PCR on antral and colonic biopsies was negative. Given the suspicion of vasculitic colitis, complement levels, Antineutrophil antibody (ANA) and c-ANCA (PR3 Ab) were assessed—complements and ANA were normal; c-ANCA titer was 162 IU/mL (positive). The patient received 500 mg methylprednisolone daily for three consecutive days and mycophenolate mofetil; she improved and was discharged on prednisolone and mycophenolate with recommendation for outpatient follow-up in two weeks. Forty-eight hours post-discharge she was readmitted with massive rectal bleeding, pallor, anemia (Hb 2.9 g/dL), hypotension requiring PICU admission. Laboratory data: WBC 13,800 (PMN 77.5 %, Lymph 19 %), Hb 2.9 g/dL, Hct 10.3 %, MCV 91.3 fL, PLT 354 000, ESR 24 mm/h, CRP 2.6 mg/L, PT/INR/PTT normal, liver function tests (LFTs) normal; BUN 48 mg/dL, SCr 5.1 mg/dL; Na, K normal; Ca 8.1 mg/dL, P 3.6 mg/dL, Albumin 2.4 g/dL. She received normal saline, blood transfusion and after stabilization a small bowel RBC scan and a Meckel's diverticulum scan were performed showing active focal bleeding site in the ileum (compatible with SMA territory) (Figure 1). CT angiography of abdominal vessels revealed contrast extravasation into ileal loops and prominent intramural vessels adjacent to extravasation, suggestive of vascular malformation. During angiography multiple microaneurysms were identified throughout the small intestine, especially in the ileum and colon; embolization was deemed unfeasible. Aggressive therapy for GPA relapse was instituted: five sessions of plasmapheresis, 2 gr/kg intravenous immunoglobulin (IVIG), and 375mg/m^2 rituximab (Table 1). The bleeding ceased, and the patient was discharged 12 days later with good condition and with no further drop in hemoglobin. At subsequent follow-up she remained stable without GI bleeding or Hb decline.

DISCUSSION

This case underscores a highly unusual presentation of GPA in a dialysis-dependent child with GI bleeding and diffuse small-bowel microaneurysms, despite her vasculitis remaining inactive for at least one year following dialysis. Although gastrointestinal involvement in GPA is rare, particularly in pediatric populations, it poses a significant life-threatening risk. GI vasculitis in GPA is reported more often in males, although documented cases are scarce.[1]

Patients with GPA are generally advised to continue immunosuppressive medications for at least two years to prevent relapse. However, in ESKD patients undergoing maintenance dialysis, the relapse rates of ANCA-associated vasculitis appear to decrease; i.e., in one study, relapse rate dropped from 0.20 episodes/person-year pre-dialysis to 0.08/person-year post-dialysis (P = .0012).[2-4] The very low relapse rate in the ESKD population complicates the assessment of the independent effect of continued maintenance immunosuppression. On the other hand, immunosuppressed patients with ESKD experience an infection risk that is about twice as high as that of individuals not undergoing such treatment. Hence, some authors recommend restricting immunosuppression to those with active

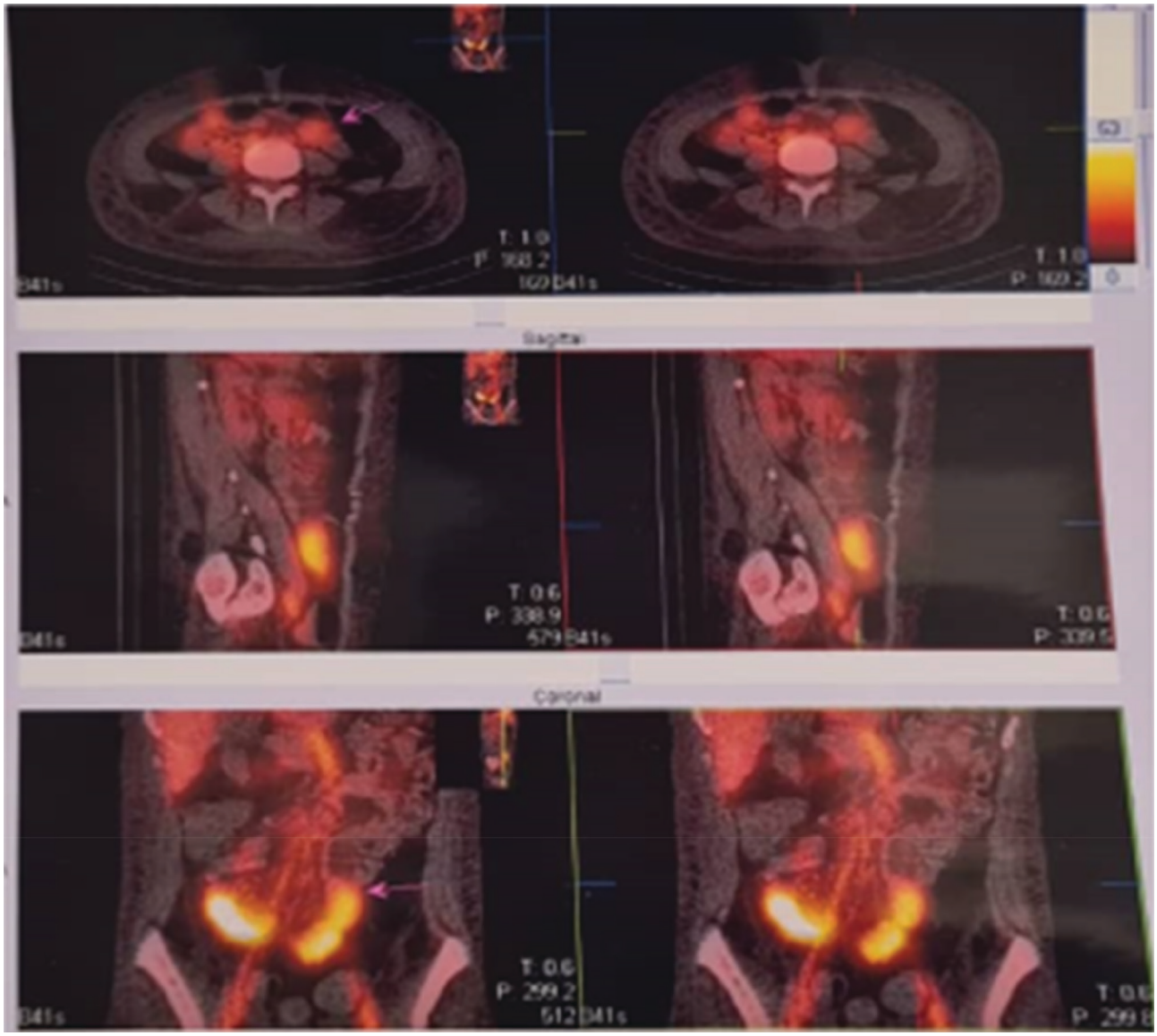

Figure 1. Small bowls and Meckel's Diverticula RBC Scan

Table 1. Case Treatment Summary (GPA Flare with Chronic Kidney Failure)

Treatment Steps	Medication/Procedure	Dose / Duration
Initial high-dose corticosteroid therapy	methylprednisolone pulse	500 mg IV daily for three consecutive days
Adjunctive therapy	Plasma exchange	five sessions
Immunomodulatory therapy	IVIG	2 gr/kg
Biologic therapy	Rituximab	Single dose (375mg/m^2)

extrarenal vasculitis manifestations.[5] Despite the historically lower relapse risk in dialysis patients, this case demonstrates that relapse remains possible and may present atypically. Therefore, decisions to discontinue maintenance immunosuppressive therapy should be made cautiously, with close monitoring

CONCLUSION

Relapsing ANCA-associated vasculitis in hemodialysis patients presents a unique challenge, while relapse risk in hemodialysis patients is low, especially in MPO-ANCA positive cases, and maintenance immunosuppressive therapy may be discontinued earlier than in non-hemodialysis

patients, the risk remains non-negligible. Clinicians must carefully evaluate the risks of continuing immunosuppression (notably infection in ESKD) against the possibility of relapse, especially in the presence of extrarenal symptoms.

CONSENT

Written informed consent was obtained from the patient's guardian for publication of this case report and any accompanying images.

ETHICAL APPROVAL

This study was approved by the Ethics Committee of Iran of University of Medical Sciences.

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DECLARATIONS OF COMPETING

The authors declare no conflict of interest.

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