OXFORD

Oxford Medical Case Reports, 2023, **11**, 430–432
https://doi.org/10.1093/omcr/omad127
Case Report

Intestinal perforation as a first presentation of granulomatosis with polyangiitis: unusual case report

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Abstract

Introduction: Granulomatosis with polyangiitis (GPA) vasculitis typically involves upper and lower airways and kidneys. Gastrointestinal involvement is rare, clinically reported as esophageal involvement, gastrointestinal hemorrhage, intestinal perforation, colitis, and pancreatitis. Case presentation: We present a 36 old man, with intestinal perforation, laterally diagnosed as granulomatosis with polyangiitis. Discussion: Only a few cases of intestinal perforation have been reported in the medical literature. GI symptoms may be present after the disease diagnosis in years. Intestinal perforation usually required surgery. The frequent kidney involvement of GPA is rapidly progressive glomerulonephritis, presented as acute kidney injury, usually accompanied by GI symptoms. Cyclophosphamide plus corticosteroids remain the effective therapy. The patient with GPA had a normal life expectancy due to the advances in treatment. Renal involvement and GI manifestations are considered bad prognosis predictors. Conclusion: This case report illustrates the need to consider intestinal perforation in patients with granulomatosis with polyangiitis, early surgical intervention and appropriate immunosuppressive therapy can be lifesaving.

INTRODUCTION

Granulomatosis with polyangiitis is a multisystem necrotizing small-medium vessel necrotizing vasculitis. The disease typically involves the upper and lower respiratory tracts and kidneys [1]. Gastrointestinal involvement is rare, seen in only 5%–10% of the cases, and is a poor prognostic factor. Both small and large bowel can be affected, leading to life-threatening complications [1, 2].

We present multiple intestinal perforations in a patient, laterally diagnosed as Granulomatosis with polyangiitis, with glomerulonephritis.

CASE REPORT

A 36 years old man came to the emergency department at Al-Assad University Hospital in Damascus, Syria, with severe colic abdominal pain of two months duration, no nausea, vomiting, or changes in bowel habits. The pain was generalized with no triggers and no relation to meals, usually resolves by taking painkillers, it became consistent and unbearable 2 days before admission. The patient mentioned that he had psoriasis, confirmed by biopsy, at the age of 30 years old, treated with topical steroids, and ultra-violet. No other previous medical history of the family was positive.

Physical examination showed that blood pressure was 110/70 mm/Hg, heart rate was 100/min, respiratory rate was 18/min and temperature was 37 degrees by mouth.

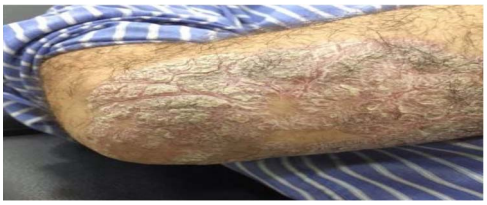

Figure 1. Multiple erythematic lesions.

We recognized red eyes, there were multiple erythematic lesions with elbows, and knees (Figure 1).

Abdominal examination showed tenderness and tough all over the abdomen, liver span was 10 c.m, and spleen was not palpable, rectal examination was normal.

Ophthalmological examination found an anterior uveitis, steroid drops, and pupil dilators were prescribed.

Laboratory findings were: Hematocrit 24%, white blood cells 13.7/10 mm3 (75% neutrophils and 20% lymphocyte), platelets 962 000 mm3, erythrocyte sedimentation rate 65 mm/h1, urea 30 mg/dl, and creatinine 0.75 mg/dl. Urinalysis was normal on admission.

A blood smear revealed a very high number of platelets, a bone biopsy was unremarkable consequently, and it was considered a secondary thrombocytosis.

Abdominal X-ray revealed air under the right and left diaphragm.

Received: May 29, 2023. **Revised:** July 13, 2023

Table 1.

Laboratory tests	Admission	After 7 days
Creatinine	0.3 (up to 0.8 mg/dl)	11.8 mg/dl
Urea	10 (up to 30 mg/dl)	80 mg/dl
Protein of Urine 24 h	–	3332 mg/24 h
Potassium	2.5 (3.5–5 mg/dl)	5.5 mg/dl

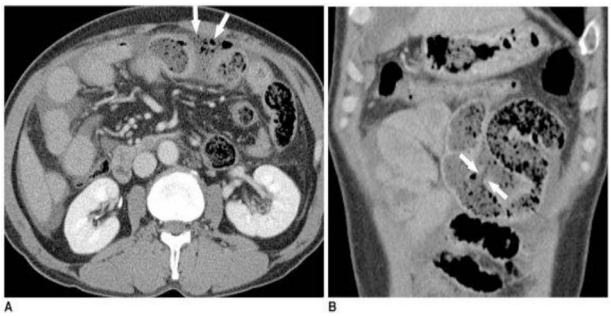

Figure 2. CT scan.

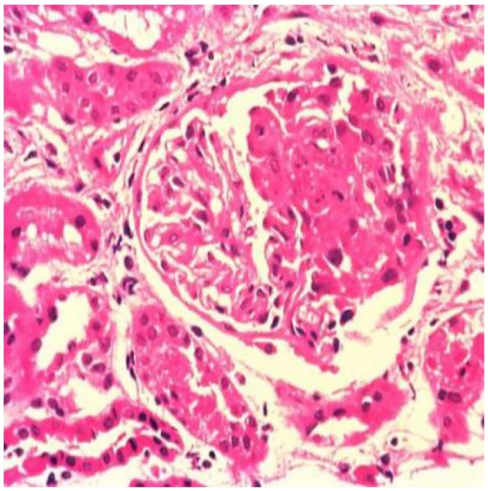

Figure 3. Renal biopsy.

Computed Tomography showed concentrated free air (arrows) in the jejunal loop, and demonstrates discontinuity of the jejunal wall (arrows), (Fig. 2).

An urgent laparotomy was done and revealed perforations, the surgeons sutured the perforations.

Seven days after the operation, the creatinine level increased to 11.8 mg/dl (up to 0.8 mg/dl), with the appearance of protein urea in urinalysis. 24 h protein was 3332 mg/dl, and complement levels were normal (Table 1).

The immune profile showed that C-ANCA in ELISA tests was positive, at a level of 320 u/ml (negative: ≤19 AU/ml. Equivocal: 20–25 AU/ml. Positive: ≥26 AU/ml).

A renal biopsy was performed, which revealed focal necrotizing glomerulonephritis, with negative immunofluorescent compatible with pauci- immune vacuities (Figs 3 and 4).

Hemodialysis was started. The induction therapy consisted of methylprednisolone intravenously (1 g/day 5 days), followed by oral prednisone (8omk/day), and cyclophosphamide (0.75 g/m2), intravenously once a month for 6 months. The patient improved rapidly, protein-urea disappeared, and creatinine levels returned to the normal limits after 2 months. At that time, he was taking 50 mg/day predlone.

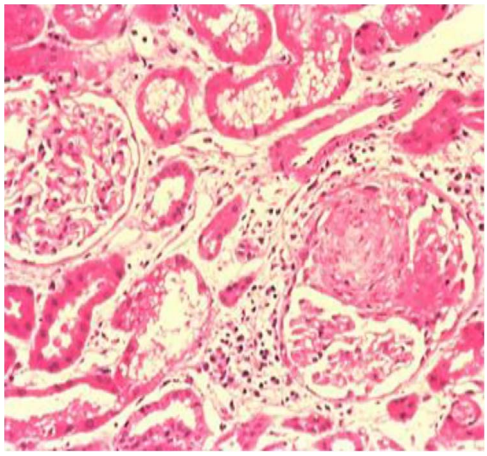

Figure 4. Renal biopsy.

DISCUSSION

Gastrointestinal involvement in granulomatosis with polyangiitis is usually rare, ranging from 5% to 10% [1, 2]. GI manifestations were clinically reported as enter colitis, gastrointestinal hemorrhage, intestinal perforation, esophageal involvement, and recurrent pancreatitis [3, 4].

Only a few cases of intestinal perforation have been reported in the medical literature. The mean age at onset of the first symptoms of vasculitis in all mentioned cases, was more than 40 years old, while our patient was 36 years. On the other hand, colitis, as an unusual presentation of Wegener's granulomatosis, was mentioned by Sinnott JD, et al [5].

GI symptoms may be present after the disease diagnosis in years, occurring during relapse of the vasculitis [3], while it was the first manifestation in our case.

The intestinal perforation and necrosis should be regarded as a complication of granulomatosis with polyangiitis itself, rather than medical therapy [6], which is compatible with our case, as the patient did not receive any immunosuppressants [1, 2].

Intestinal perforation usually required surgery [1, 2], as in our case.

The frequent kidney involvement of GPA (RPGN) is rapidly progressive glomerulonephritis, leading to chronic kidney disease, and end-stage renal disease. Renal involvement is presented in only 10%–20% of cases, and in 80% of patients within two years of disease onset, which is incompatible with our case. RPGN is characterized clinically by a rapid decrease in the glomerular filtration rate, and histologically pauci-immune necrotizing and extensive glomerular crescent formation [7, 8], as in our case.

At present, despite the increasingly wide range of potential therapies, cyclophosphamide plus corticosteroids remain the effective therapy for inducing and sustaining remission. Although, therapy biologic treatment such as rituximab might be a treatment option [1, 2]. Our patients were treated with cyclophosphamide plus corticosteroids.

The 10-year survival rate is 40% with kidney injury, and 60% to 70% without renal involvement, in GPA patients, and may have a normal life expectancy due to the advances in treatment [9]. The presence of dialysis-requiring renal damage is the most important risk factor for mortality in GPA patients. These patients should be followed more closely and carefully to improve survival. In addition to irreversible organ dysfunction, or due to the consequences of treatment [1, 2]. Unfortunately, our young patient had renal involvement, which rapidly progress and require dialysis, in addition to the GI presentation. There is a high incidence of laparotomy and death, associated with reported cases of intestinal involvement [1, 2, 10]. Our patient needed surgery.

However, to the best of our knowledge, this is the first case that describes the coexistence of GPA, and psoriasis, and the second case presented with intestinal perforation, as a first manifestation of GPA, and also needed dialysis and cyclophosphamide to improve.

CONCLUSIONS

This case report illustrates the need to consider intestinal perforation in patients with granulomatosis with polyangiitis, early surgical intervention, and appropriate immunosuppressive therapy can be lifesaving.

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