Polyarteritis nodosa and acute abdomen: A role for laparoscopy?

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A B S T R A C T
Mesenteric vasculitis secondary to polyarteritis nodosa represents an atypical but potentially life-threatening cause of bowel ischemia and acute abdomen. The patient presented with severe abdominal pain of recent onset, pitting edema of the legs, renal failure and bowel wall thickening suggestive of mesenteric ischemia on CT scan. Early laparoscopy allowed to rule out proximal bowel necrosis and resection was avoided. The patient was successfully managed with corticosteroid therapy and repeated hemodialysis sessions.

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1. Introduction

Acute mesenteric ischemia generally represents a surgical emergency and it may occur as a result of arterial or venous occlusion, nonocclusive mesenteric hypoperfusion, or a systemic vasculitis. The precise cause is difficult to ascertain even at laparotomy. Polyarteritis nodosa may rarely cause bowel ischemia with histological evidence of fibrinoid necrosis of the vessel wall [1]. We report a case of polyarteritis nodosa presenting with a clinical picture of acute abdominal pain, pitting edema of the legs, and renal dysfunction.

2. Case report

A 57-year-old Caucasian man with a history of rheumatoid arthritis, poorly controlled arterial hypertension, and transient ischemic attack, was admitted to the Emergency Department for severe abdominal pain of recent onset. Initially he appeared slightly distressed; his temperature was 36.9°C, blood pressure 120/80 mmHg, pulse 120 beats/min, and oxygen saturation 96% on room air. On physical examination, generalized abdominal guarding and pitting edema of the legs with pectechiae was noted (Fig. 1). The laboratory findings were remarkable for elevated white blood cell count (20.960/μL), C-reactive protein (19.9 mg/dL), and impaired renal function (GFR 48 mL/min/1.73 m², creatinine 1.85 mg/dL, urea 97 mg/dL). Abdominal computed tomography with contrast showed diffuse thickening and edema of the proximal small bowel loops suggestive of mesenteric ischemia (Fig. 2). Wide-spectrum antibiotic therapy was initiated and minimally invasive surgical exploration was planned. At laparoscopy, an edematous jejunal loop without evidence of perforation was found 20 cm distal to the Treitz's ligament. No resection was performed. The postoperative course was complicated by fever and worsening of the pitting edema of the legs; there was also a further increase of the inflammatory markers and creatinine levels (3.91 mg/dL) with the appearance of proteinuria (250 mg/dL) and hematuria (1 mg/dL). Since the clinical picture was consistent with the diagnosis of polyarteritis nodosa, corticosteroid therapy (Prednisone 75 mg/day) was initiated and multiple sessions of hemodialysis were performed. Serology was negative for hepatitis B and C virus, anti-Beta2-glycoprotein antibodies, anti-nuclear antibodies, anti-cardiolipin antibodies, anti-citrulline antibodies, anti-native DNA autoantibodies, cytoplasmic-Anti-Neutrophil Cytoplasmic Antibodies (ANCA), perinuclear-ANCA, and rheumatoid factor. Repeat abdominal computed tomography with contrast showed kidney hypoperfusion and persistent small bowel edema. Over the following days the pitting edema of the legs decreased and the renal function improved. A skin biopsy eventually confirmed the diagnosis of polyarteritis nodosa.

3. Discussion

Vasculitis, including polyarteritis nodosa, represent an atypical but potentially life-threatening cause of acute abdomen. Polyarteritis nodosa is a systemic necrotizing vasculitis that typically affects
small to medium size arteries. Gastrointestinal involvement with mesenteric ischemia is relatively common in these patients and may be life-threatening due to diagnostic delay or inappropriate management. Although mesenteric vasculitis is a rare cause of intestinal infarction, representing about 2% of cases [2], it appears from our review of the literature that 38.3% of patients with polyarteritis nodosa present with gastrointestinal manifestations of the disease; of these individuals, 42.9% underwent emergency surgery via laparotomy with an exceedingly high operative mortality rate (Table 1) [3–11].

Table 1
Prevalence and mortality rates of gastrointestinal (GI) manifestations in patients with polyarteritis nodosa (literature review).

|                  | No. patients | No. patients with GI manifestations | No. patients undergoing emergency surgery | Operative mortality |
|------------------|--------------|------------------------------------|------------------------------------------|---------------------|
| Cohen et al. [3] | 53           | 14                                 | 8                                        | 6 (7.5%)            |
| Ito et al. [4]   | 2            | 2                                  | 2                                        | 0 (0%)              |
| Damani et al. [5]| 1            | 1                                  | 1                                        | 1 (100%)            |
| Levine et al. [7]| 54           | 24                                 | 13                                       | 3 (23%)             |
| Kariv et al. [6] | 1            | 1                                  | 1                                        | 0 (0%)              |
| Gonzalez-Gay et al. [8]| 1            | 1                                  | 1                                        | 0 (0%)              |
| Pagnozzi et al. [9]| 348        | 132                                | 48                                       | 30 (62.5%)          |
| Hiraie et al. [10]| 1           | 1                                  | 1                                        | 1 (100%)            |
| Shirai et al. [11]| 462         | 177/462 (38.3%)                   | 76/177 (42.9%)                           | 41/76 (53.9%)       |

Acute onset abdominal pain accompanied by pitting edema of the legs and impaired renal function should always remind the clinical diagnosis of polyarteritis nodosa and possible occlusive mesenteric vasculitis with or without bowel infarction. No specific diagnostic marker has ever been identified, and the diagnosis of polyarteritis nodosa is most often confirmed by biopsy. Early laparoscopic exploration is safe and may play a decisive role to exclude ischemic bowel necrosis and avoid resection when clinical and CT findings are equivocal [12]. Corticosteroid therapy and hemodialysis should be first-line therapy in these patients.

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflicts of interest
Nothing to declare.

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There were no sponsors.

Ethical approval
This case report did not require ethical approval.

Consent
Written and signed fully informed consent was obtained from the patient.

Author contribution
EA and LP designed the study, ST interpreted the radiological imaging studies, LB revised the manuscript.

Guarantor
Luigi Bonavina.

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