

# Microscopic Polyangiitis Presenting with Acute Abdominal Pain Due to Rare Small Intestine Involvement

Yavuz Çağır<sup>1</sup> , İlhami Yüksel<sup>1,2</sup> 

<sup>1</sup>Department of Gastroenterology, Ankara Bilkent City Hospital, Ankara, Türkiye

<sup>2</sup>Department of Gastroenterology, Ankara Yıldırım Beyazıt University, Ankara, Türkiye

**Cite this article as:** Çağır Y, Yüksel İ. Microscopic Polyangiitis Presenting with Acute Abdominal Pain Due to Rare Small Intestine Involvement. *J Enterocolitis*. 2023;2(3):69-70.

**Corresponding author:** Yavuz Çağır, e-mail: yvzcgr@hotmail.com

**Received:** December 13, 2023 **Accepted:** December 17, 2023

DOI:10.14744/Jenterocolitis.2023.231659



Content of this journal is licensed under a Creative Commons Attribution-NonCommercial 4.0 International License.

## Abstract

A 44-year-old male patient was admitted to the emergency department with symptoms of acute abdominal pain, diarrhea, weight loss, and a rash on his legs. Abdominal computed tomography revealed wall thickening and diffuse ascites in the ileum. Colonoscopy showed focal nodular lesions and erosions in the terminal ileum. The diagnosis of microscopic polyangiitis (MPA) was established based on vasculitis symptoms, the presence of cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) in the blood, and a skin biopsy confirming leukocytoclastic vasculitis. High-dose prednisolone (1 mg/kg) was initiated to address the kidney, skin, neurological, and severe gastrointestinal system involvement. The patient made a full recovery within two weeks of treatment initiation. Herein, we report a case of MPA with an initial presentation of diffuse small intestinal involvement manifested by acute abdominal pain. To the best of our knowledge, this specific presentation has not been previously documented in medical literature.

**Keywords:** Abdominal Pain, microscopic polyangiitis, small intestinal involvement

## INTRODUCTION

Microscopic polyangiitis (MPA) is a pauci-immune necrotizing vasculitis that affects small vessels, including small arteries, arterioles, venules, and capillaries. This condition can impact various organs and systems, such as the skin, muscle, lungs, kidneys, brain, heart, eyes, peripheral nervous system, and gastrointestinal tract. There have been reports of colonic involvement in MPA confirmed by endoscopic examination, and small intestinal lesions have been identified exclusively through double-balloon endoscopic procedures. While gastrointestinal bleeding and intestinal involvement are recognized manifestations, the occurrence of small bowel complications that mimic an acute abdomen represents a rare and serious aspect of MPA. In this report, we describe the case of a patient with MPA who presented with acute abdominal pain, diffuse small intestinal involvement, and other complications, including renal, neurological, and cutaneous associations.

## CASE REPORT

A 44-year-old man presented at the emergency department with severe abdominal pain, diarrhea, weight loss, and a rash on his legs. The onset of his abdominal pain occurred 15 days prior and had gradually intensified and become more widespread. Approximately a week ago, he began experiencing non-bloody diarrhea, occurring eight times a day, and a rash developed on his legs. His medical history was unremarkable. An abdominal examination revealed tenderness, guarding, and rebound tenderness. Pretibial edema was observed, along with a purpuric rash on his legs and the dorsum of his feet. A neurological examination indicated peripheral neuropathy, which was confirmed by electromyography.

Laboratory tests showed negative results for antinuclear antibody (ANA), ANA profile, perinuclear antineutrophil cytoplasmic antibody (pANCA), and anti-glomerular basement membrane antibody (anti-GBM Ab), while cytoplasmic ANCA (cANCA) was positive (23.160 U/ml). Abdominal computed tomography (CT) scans revealed wall thickening of the small intestine and diffuse ascites. Colonoscopic examination identified focal nodular lesions and erosions in the terminal ileum, but the colon appeared normal. An ileal biopsy ruled out inflammatory bowel disease. Skin biopsy findings were consistent with leukocytoclastic vasculitis. With the diagnosis of ANCA-positive microscopic polyangiitis (MPA) and the presence of renal, cutaneous, neurological, and severe gastrointestinal tract involvement, treatment with high-dose prednisolone (1 mg/kg) was initiated. The patient fully recovered within two weeks.

## DISCUSSION

Microscopic polyangiitis (MPA) is a multisystemic, antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis frequently affecting the kidneys and lungs. Though less commonly implicated, the gastrointestinal tract and other organ systems can be involved. The predominant gastrointestinal symptom in MPA is abdominal pain, followed by diarrhea, vomiting, and bleeding. Abdominal pain is observed in 97% of patients, making it an almost universal finding. Consequently, while subclinical gastrointestinal involvement may be detected through systematic abdominal

examinations like CT or angiography, the absence of abdominal pain generally excludes the diagnosis of gastrointestinal tract vasculitis.<sup>1</sup> In our patient, acute abdominal pain and subsequent diarrhea were initial clinical indications of the disease, representing some of the rarer symptoms that could suggest a diagnosis of MPA.

The patient's clinical presentation, including gastrointestinal, renal, and neurological involvement, along with characteristic skin signs, indicated vasculitis affecting small vessels. A skin biopsy from suspected vasculitic lesions confirmed leukocytoclastic vasculitis, and cANCA positivity further supported the MPA diagnosis.

Differential diagnoses for MPA include polyarteritis nodosa, other ANCA-associated vasculitides, connective tissue diseases like systemic lupus erythematosus and rheumatoid arthritis, and anaphylactoid purpura. The term "ANCA-related vasculitis" encompasses Granulomatosis with polyangiitis (formerly known as Wegener's granulomatosis) and MPA. Reports suggest pANCA positivity in 50 to 80% of MPA cases and about 10% in Wegener's granulomatosis, whereas cANCA is positive in 70 to 90% of Wegener's granulomatosis and approximately 10% of MPA. Wegener's granulomatosis was excluded in this patient due to the absence of granulomas in the biopsy and the lack of nasal polyps, sinusitis, or lung nodules.<sup>1</sup> Our case was marked by cANCA positivity, an atypical finding in MPA, which typically exhibits pANCA positivity.

While gastrointestinal involvement in MPA is not uncommon, diagnosing it via endoscopic biopsy is challenging due to the small size and superficial nature of the samples, which affect small vessels. Even in the absence of histological evidence, treatment should be initiated when clinical suspicion for vasculitis is high.<sup>2</sup> Prompt treatment of gastrointestinal manifestations in ANCA vasculitis is crucial, as confirmation of diagnosis can be arduous and time-consuming. Immunosuppressive therapy has significantly improved the prognosis of necrotizing vasculitis. Given the severity of the condition, delaying treatment for a tissue diagnosis is often not feasible.<sup>3-5</sup> Therefore, in complex cases like our patient, it is imperative to swiftly consider

differential diagnoses and commence appropriate immunosuppressive treatment.

## CONCLUSION

To date, there have been no reports of microscopic polyangiitis (MPA) presenting with small intestinal involvement manifesting as acute abdominal pain. We have described a patient with MPA who presented with multisystemic involvement, including acute abdominal pain. It is essential for physicians to consider that MPA, although rare, can significantly affect the small intestine and present with serious gastrointestinal symptoms.

**Informed Consent:** Written informed consent was obtained from the patient who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Data Collection and/or Processing - Y.Ç.; Writing - Y.Ç., İ.Y.; Critical Review - İ.Y.

**Declaration of Interests:** The authors declare that they have no competing interest.

**Funding:** The authors declare that this study has received no financial support.

## REFERENCES

1. Eriksson P, Segelmark M, Hallböök O Frequency, Diagnosis, Treatment, and Outcome of Gastrointestinal Disease in Granulomatosis with Polyangiitis and Microscopic Polyangiitis. *J Rheumatol.* 2018;45(4):529-537. [\[CrossRef\]](#)
2. Watts R, Lane S, Hanslik T, et al. Development and validation of a consensus methodology for the classification of the ANCA-associated vasculitides and polyarteritis nodosa for epidemiological studies. *Ann Rheum Dis.* 2007;66(2):222-227. [\[CrossRef\]](#)
3. Holstein A, Titz U, Maschuw K. Intestinal manifestation of polyangiitis with massive bleeding. *Dtsch Arztebl Int.* 2019;116(16):288. [\[CrossRef\]](#)
4. Rašić S, Džemidžić J, Karasalihović Z, Herenda V, Rebić D. Microscopic polyangiitis presented with polyneuropathy of lower extremities and ANCA-associated glomerulonephritis: Case Report. *Bosn J Basic Med Sci.* 2012;12(1):55-58. [\[CrossRef\]](#)
5. Segraves JM, Iyer VN. Microscopic polyangiitis: atypical presentation with extensive small bowel necrosis, diffuse alveolar hemorrhage, and renal failure. *Respir Med Case Rep.* 2017;21:12-15. [\[CrossRef\]](#)