Recurrent Asymptomatic Sigmoid Diverticular Perforation in a Patient with Pemphigus Vulgaris on Immunosuppressive Therapy: A Case Report

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Patient: Male, 57
Final Diagnosis: Perforated diverticuli
Symptoms: Asymptomatic
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare co-existence of disease or pathology
Background: Perforation of the colon is associated with high mortality and requires early diagnosis. However, the diagnosis of perforation from atypical causes can be a diagnostic challenge. This report is of a rare case of recurrent sigmoid colonic perforation in a patient with diverticular disease who did not present with an acute abdomen but who had pemphigus vulgaris treated with immunosuppressive therapy.

Case Report: A 57-year-old man with pemphigus vulgaris was treated with steroids, non-steroidal anti-inflammatory drugs (NSAIDS), and azathioprine. He had episodes of abdominal bloating but denied any other symptoms. He was diagnosed with spontaneous sigmoid diverticular perforation without presenting with an acute abdomen.

Conclusions: Diverticular perforation can be asymptomatic in patients on immunosuppressive therapy. Therefore, there should be a high index of suspicion for bowel perforation in patients with abdominal symptoms who are treated for skin diseases, such as pemphigus vulgaris, and are on steroids and other immunosuppressive treatments.

MeSH Keywords: Diverticulitis • Intestinal Perforation • Pemphigus

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Background

Large bowel perforation is rare, and is mainly caused by colorectal malignancy and diverticulitis, and can be associated with severe complications. Colonic perforation is associated with high mortality with a rate that ranges from between 16.9–19.6% [1,2]. The surgical approach to the management of large bowel perforation is controversial. Large bowel perforation may be immediately suspected when the patient presents with an acute surgical abdomen. However, it can be a diagnostic challenge when the clinical presentation is atypical. There have been few previously reported cases with a presentation of recurrent colonic perforation without any acute abdominal signs.

This report is of a rare case of recurrent sigmoid colonic perforation in a patient with diverticular disease who did not present with an acute abdomen but who had pemphigus vulgaris treated with immunosuppressive therapy. This case highlights the importance of ensuring that patients taking multiple immunosuppressive drugs who present with nonspecific abdominal signs are thoroughly investigated to avoid delays in definitive management.

Case Report

A 57-year-old man was admitted to the emergency room (ER) with fever, dysuria, and left flank pain. His past medical history was significant for pemphigus vulgaris, diabetes mellitus, deep vein thrombosis of the right lower limb complicated by pulmonary embolism, and chronic hydronephrosis with an atrophic left kidney. His medications on admission included enoxaparin sodium, metformin, gliclazide, ibuprofen, prednisone 100 mg, and azathioprine. On physical examination, he appeared unwell and abdominal palpation identified left costophrenic angle tenderness. His abdomen was otherwise soft, non-distended, and non-tender. Laboratory investigations showed a white blood cell (WBC) count that was within the normal range. Urine culture identified antibiotic resistant *Escherichia coli*. This patient was diagnosed with pyelonephritis and was hospitalized to receive intravenous antibiotic treatment.

On the fifth day following hospital admission, the patient began to complain of abdominal bloating (Figure 1) and constipation. Abdominal distension was partially relieved by defecation. He denied any pain. On physical examination, the abdomen was still soft and non-tender but was distended. His vital signs were within the normal range. Auscultation identified hypoactive bowel sounds. These symptoms were attributed to prolonged recumbency, and so increased ambulation was advised and laxatives were prescribed. His WBC count was still within the normal range.

On the seventh day following hospital admission, the patient’s clinical condition had not improved. An abdominopelvic computed tomography (CT) scan was performed, which showed perforated sigmoid diverticulitis with peri-sigmoidal abscesses (Figure 2) The imaging findings were consistent with a Hinchley III perforation associated with purulent peritonitis due to diverticulitis of the colon. Therefore, laparoscopic drainage of the purulent peritonitis and an abscess cavity was performed (Figure 3). The patient’s condition improved, and there were no complications following laparoscopic drainage.
Pemphigus vulgaris is an IgG-mediated autoimmune disease that causes blisters and erosions of the skin and oral mucosa through acantholysis of the stratified squamous epithelium [3]. Systemic corticosteroids are used as first-line treatment for pemphigus vulgaris, but azathioprine and mycophenolate are also commonly prescribed in conjunction with steroids to reduce the steroid dose and the adverse effects. Recently, rituximab and plasmapheresis have been used in refractory cases, usually in conjunction with cytotoxic therapy [4].

Colon diverticulosis is the presence of multiple uninflamed diverticula (pouches) in the colon and is usually asymptomatic. However, symptoms can range from mild abdominal pain, usually in the left lower quadrant, to severe generalized peritonitis and shock [5]. Corticosteroid use has previously been described as a cause of spontaneous diverticular perforation [6–8]. The immunosuppressive effect of corticosteroids results in the impaired ability to contain the perforation during the early stages [9]. Also, the use of non-steroidal anti-inflammatory drugs (NSAIDs) in patients with diverticular disease is associated with an increased risk of perforation mainly through damage to epithelial cells, reduced mucin secretion [9], and reduced synthesis of prostacyclin [10]. The patient presented in this case report had been treated with both corticosteroids and NSAIDs, which increased the risk of diverticular perforation.

This patient had an unusual presentation with two consecutive episodes of perforated diverticula associated with abdominal distension. This unusual presentation resulted in a delay in diagnosis and definitive management. To our knowledge, there have been no previously reported cases with a similar presentation. A previously published report of a symptomatic case in a patient with rheumatoid arthritis attributed spontaneous diverticular perforation to the use of the disease-modifying antirheumatic drug (DMARD), methotrexate [11]. However, there have been no subsequent studies that have confirmed an association between the use of DMARDs and spontaneous diverticular perforation. In this patient, immunosuppressive effects of both steroids and the DMARD, azathioprine, may have completely abolished the signs of peritonitis or inflammation, resulting in the lack of symptoms on presentation.

The aim of presenting this case report was to highlight the importance of the medical history, including ongoing treatment for other diseases, when evaluating patients who present acutely to hospital. Immunosuppressive medication may mask the signs of inflammation and reduce symptoms, including those usually associated with perforation of a viscus, or the symptoms of an acute surgical abdomen, and may delay definitive treatment. In patients who are immunosuppressed investigations, including imaging, should be performed as an
emergency to prevent delay in definitive management. A further point to stress is that patients who are taking immunosuppressive medications also have impaired wound healing, which may alter to approach to surgical management of colonic perforation in diverticulitis, for example with a more aggressive approach that includes Hartmann’s procedure instead of laparoscopic lavage.

**Conclusions**

Diverticular perforation can be asymptomatic in patients on immunosuppressive therapy. Therefore, there should be a high index of suspicion for bowel perforation in patients with abdominal symptoms who are treated for skin diseases, such as pemphigus vulgaris, and are on steroids and other immunosuppressive treatments.

**Conflict of interest**

None.

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