

CASE NOTE

An unexpected severe complication after a negative laparoscopic appendectomy

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CASE REPORT

A 67-year-old woman with a history of myelodysplastic syndrome (MDS) was on holiday in another country when abdominal pain developed. Her MDS had been stable over the previous 2 decades, and her outpatient monitoring had been cancelled. She went to the local emergency department with pain in the right lower quadrant. Laboratory results showed elevated C-reactive protein (CRP) and white blood cell (WBC) counts. The patient underwent a diagnostic laparoscopy for suspected acute appendicitis. At laparoscopy, her appendix was not inflamed and there was no other cause for her abdominal pain. Postoperatively, what at first looked like a wound infection developed at the trocar opening in lower left abdomen. The wound turned necrotic. Surgeons performed a necroectomy of the wound and placed a central venous catheter in her left subclavian vein. The patient was started on antibiotics. Since she was on holiday in a foreign hospital, she requested a transfer to a hospital in her home country. Arrangements were made to transfer the patient to our ward.

On admission, she had a fever (39°C), elevated CRP (3038.16 nmol/L) and WBC counts ($14.7 \times 10^9/L$) and refractory anemia secondary to myelodysplastic syndrome. She had a large wound in her lower left abdomen with some debris (Fig. 1A). She also had thoracic pain at the site where the central venous catheter had been placed; the former access site was slightly red and swollen. A skin incision for suspected wound infection did not reveal any pus. The next day, she became clinically septic and was admitted to our intensive care unit. A thoracic and abdominal computed tomography (CT) scan revealed no focus



Fig. 1. Photographs show (A) the wound in lower left abdomen 2 weeks after laparoscopic appendectomy and (B) the wound where the central venous access point used to be. The wound has a typical aspect of pyoderma gangrenosum characterized by ulcerations with violaceous, undermined borders. We obtained 2 biopsy specimens and diagnosed pyoderma gangrenosum.

for her sepsis. The initial diagnosis was septic shock caused by wound infection in an immune-compromised state. The antibiotic therapy was not effective, and the lesion at the venous access point worsened over the next couple of days (Fig. 1B). Blood cultures, a throat swab and ulcer fluid yielded no micro-organisms. Finally, a biopsy and culture of the skin lesion showed severe infiltration of neutrophils in the dermis, without any evidence of infection (Fig. 2). We diagnosed the lesion as pyoderma gangrenosum and began systemic administration of corticosteroids. The skin lesion responded to the therapy, and her CRP level normalized. We discharged the patient when the lesions were almost healed. The prednisolone administration was tapered after 6 months of maintenance therapy, and she experienced no recurrence of pyoderma gangrenosum.

DISCUSSION

Pyoderma gangrenosum (PG) was first described by Brunsting and colleagues in 1930¹ in a cohort of patients with inflammatory bowel disease (IBD). The typical lesions of PG are characterized by ulcerations with violaceous, undermined borders. An inflammatory areola of erythema may surround the ulcer border during the acute phase, and the patient typically experiences severe pain. The lesions classically develop in patients with IBD across the pretibial or peristomal skin and are well-known to surgeons who treat these patients. Still, PG in patients with IBD is relatively rare. A study involving 1043 patients with IBD reported an incidence of 0.6%.² Pyoderma gangrenosum is a neutrophilic dermatosis histologically characterized by dermal neutrophilic infiltrates with no infectious or other identifiable etiology.³ The disease may be associated with a

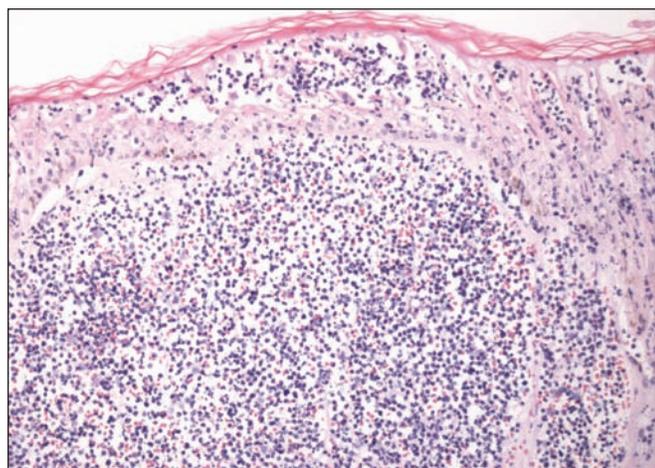


Fig. 2. Histopathology, using hematoxylin and eosin staining, revealed a severe infiltration of neutrophils in the dermis, without any evidence of infection (original magnification $\times 40$).

variety of other systemic disorders such as rheumatoid arthritis and various hematologic malignancies. Only 31 cases of PG in patients with myelodysplastic syndrome have been reported in the literature.⁴ The differential diagnoses of PG include bacterial cellulitis, atypical mycobacterial infections and deep fungal mycoses, including blastomycosis, sporotrichosis and cryptococcosis.³ The cause of PG is still debated. Many patients can relate the development of the skin lesions to recent trauma to the affected area, a phenomenon known as pathergy. Using the principle of pathergy, it has been suggested that minor trauma to the skin such as maintenance of stoma collection devices may initiate the development of PG. In our patient's case, PG also developed in the trocar wound after surgery and in the subclavian venous access site after an incision was made.

Pyoderma gangrenosum is a potentially lethal disease with a mortality of 30% in some series.³ In general, the initial therapy for PG is the systemic administration of prednisolone.³ Topical treatment is insufficient, especially in patients with associated hematologic malignancies. Up to 50% of patients with PG have been reported to experience relapses.³ However, our patient did not experience a recurrence. More than 50% of patients with PG have associated systemic diseases. Ulcerative colitis is the disease most frequently found to be associated with PG, followed by Crohn disease and inflammatory arthritis. About 7% of PG cases are associated with cancer;⁵ 1% occur with hematologic malignancies, including leukemia, polycythemia vera, myelofibrosis, essential thrombocythemia and multiple myeloma. Acute myeloid leukemia and chronic myeloid leukemia are the most frequent of the hematologic malignancies associated with PG. Myelodysplastic syndrome is not often seen to be associated with PG; however, we eventually reached the correct diagnosis in our patient, and the use of high-dose pulse methylprednisolone and the long-term maintenance therapy with prednisolone were quite successful.

Competing interests: None declared.

References

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