Unique case of non-neutropaenic typhlitis in an immunosuppressed liver transplant patient

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DESCRIPTION

Typhlitis (also known as typhlenteritis or neutropaenic enterocolitis) is a recognised complication of immunosuppression following solid organ transplantation with tacrolimus increasingly thought to be a significant risk factor. It is characterised by transmural inflammation of the caecum and terminal ileum and is classically described as a triad of abdominal pain, fever and neutropaenia. The causative organism is often a Gram-positive enterococcus and typically affects immunosuppressed patients such as those undergoing neoplastic chemotherapy for haematological malignancies, postorgan transplantation or in patients with acute leukaemia or AIDS.

We describe the case of a 63-year-old woman presenting to the emergency department following two episodes of collapse, peritonitis, septic shock with acute renal failure and an elevated lactate of 10. Examination revealed generalised abdominal tenderness with peritonism. Her complete blood count was as follows: White blood count (WBC) 27.1, Red cell count 3.79, Haemoglobin 113, Haematocrit 0.345, Mean cell volume 91.0, Mean corpuscular haemoglobin 29.8, platelets 267, neutrophils 21, lymphocyte 3.7, monocyte 2.4, eosinophil 0, basophil 0.1 and nucleated RBC 0.

The patient had an extensive medical and surgical history including a panproctocolectomy with ileostomy for Crohn’s disease in 1978 (two subsequent refashioning of ileostomies), recurrent biliary sepsis resulting in cholecystectomy in 2005 and primary sclerosing cholangitis with a liver transplant in 2010. She was a type 1 diabetic with gastroparesis, hypothyroidism and anaemia. Her immunosuppressive regime included prednisolone (the long-term nature of which had led to hypoadrenalism), azathioprine and tacrolimus.

Based on her presentation, a presumptive diagnosis of ischaemic bowel was made and a non-contrast (in view of her renal failure) CT scan demonstrated evidence of intramural gas in relation to loops of small bowel located in the left iliac fossa (figures 1 and 2) with marked mesenteric and portal venous gas (figure 3). In addition, marked abdominal and pelvic vascular calcification including the Superior Mesenteric Artery and some of its branches.
Images in...

proximal branches was seen. There was no evidence of bowel perforation, free gas or fluid within the abdomen.

The patient required inotropic support in addition to fluid resuscitation and was given broad-spectrum antibiotics. The decision was made to proceed to laparotomy, the findings of which included dense adhesions but with normal healthy small bowel, which was peristalsing. There was no turbid free fluid or areas of bowel wall necrosis. The patient was transferred to the intensive care unit where over the coming days managed a complete clinical recovery with intravenous antibiotics.

Several investigations were pursued to rule out sources of infection in the immune-compromised setting; however, nil of note returned from comprehensive microbiology and virology cultures. Tacrolimus, which was held on admission, was restarted after discussion with the transplant team, and she was discharged home on the 12th postoperative day.

To the authors’ knowledge, this is the first published case of non-neutropaenic typhlitis in an immunosuppressed patient with tacrolimus thought to be a significant causative factor. It is well known that glucocorticoids increase the WBC; the predominate effect is within the first 2 weeks of commencement. This patient had been on long-term low dose steroids for several years, and while this may have contributed to a rise in the neutrophils, it is very unlikely that it masked a neutropenia.

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REFERENCES

Learning points

- Neutropaenic typhlitis is a life-threatening condition classically reported as affecting the terminal ileum and caecum, but isolated cases have reported localised incidence affecting the small bowel.
- Management of neutropaenic typhlitis is controversial due to a sparsity in published papers. The authors’ view is that initial conservative measures should be instituted for all with surgery reserved for those with peritonitis.