

# A rare case of localised gastrointestinal vasculitis in a New Zealand patient

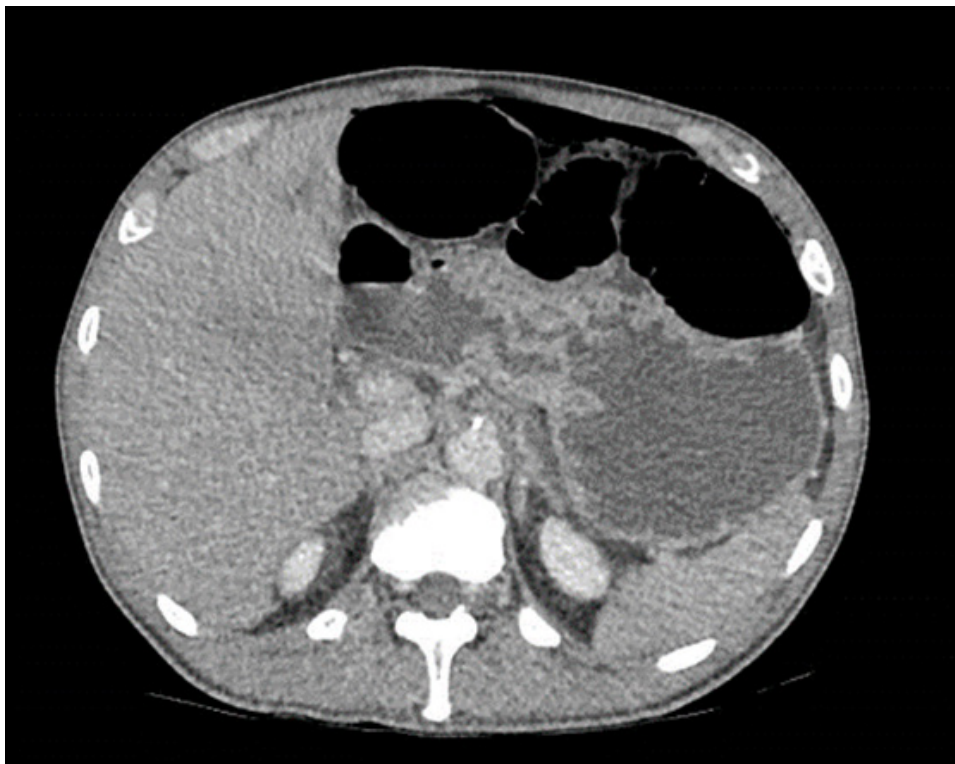
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**A** 51-year-old man with *JAK2* positive essential thrombocythaemia diagnosed a year prior, presented to Wairarapa Hospital Emergency Department with severe right sided abdominal and flank pain with a history of 3 months of similar pain. He had been discharged from Wellington Regional Hospital 5 days prior. He also presented with 13kg of weight loss over 3 months and watery diarrhoea. On physical examination he was tender to abdominal palpation and was found to be in urinary retention. An indwelling catheter was subsequently placed with moderate relief. Initial blood tests showed raised inflammatory markers (WBC  $26 \times 10^9/L$ , Neutrophils  $23 \times 10^9/L$ , CRP 94mg/L), an acute kidney injury (Creatinine 134 $\mu$ mol/L), raised platelets

( $1700 \times 10^9/L$ , which were felt to be reactive) and a normal lactate on venous blood gas. Computed tomography (CT) imaging showed non-specific dilation of the small bowel and no specific cause for his symptoms was identified. He was admitted to hospital for analgesia and observation.

Later that night, despite intravenous analgesia, the patient had further escalation of his abdominal pain. On physical examination he had developed gross peritonism and a venous blood gas revealed a lactate of 8.0mmol/L. He proceeded to an urgent CT-abdominal angiogram. This showed new free air and fluid, but with no clear point of the perforation (Figure 1). No large vessel thrombus or emboli was identified; however, it was noted that the SMA was attenuated with no filling defect.

**Figure 1:** CT-Angiography showing bowel perforation.



The patient proceeded to the acute operating theatre for an explorative laparoscopy identifying ischaemic bowel in the terminal ileum. A lower midline laparotomy incision was made, and he was found to have an ischaemic ileum. His ileum had multiple ulcerative lesions, with two areas of full thickness perforations (Figure 2). Pus and small bowel contents were seen in the abdomen and pelvis. Scattered luminal ulcerative disease was also seen in the proximal ileum and jejunum, and this region of bowel was dilated but pink and alive. The transition point between ischaemic and healthy bowel can be seen in the intraoperative photo (Figure 3). The terminal ileum was resected with staples. As the surgeon proceeded to forming a loop ileostomy, the patient became progressively hypotensive with increasing noradrenaline requirements and then became unstable in a ventricular tachycardia. Ileostomy formation was abandoned, and the bowel was left stapled in discontinuity with a nasogastric tube *in situ*.

The patient was transferred via emergency flight to Wellington. He was admitted to the intensive care unit (ICU) and reviewed by the Wellington surgical team. On arrival he was in profound septic shock with his lactate rising to 15mmol/L. Laboratory tests on arrival showed multi-organ failure with further acute kidney and now liver injury (Creatinine 250µmol/L, ALT 5722U/L). A repeat CT scan with arterial contrast showed multiple ischaemic infarcts with no arterial thrombus or embolism, suspected to be from hypoperfusion. The Wellington surgical team decided to proceed for an urgent re-look explorative laparotomy. This operation started 12 hours after the initial laparotomy. Exploration revealed extensive solid organ, large bowel and small bowel ischaemia. The surgical team deemed this ischaemia unsurvivable, and the abdomen was closed. Upon returning to the ICU, the operative findings were discussed in a meeting with his whanau, the ICU team and the surgical team. He was then extubated and died later that night.

Within the 3 months leading up to his death, this patient had undergone extensive work-up for his abdominal symptoms. These symptoms initially began with upper abdominal pain, diarrhoea and weight loss. He visited his GP twice; at first he was given a trial of oral antibiotics, and then on the second occasion was referred to the emergency department (ED). In ED he was clinically evaluated with blood tests and a CT scan. The only remarkable finding was a raised lipase

(418U/L) which was felt to be non-diagnostic. He was discharged and referred to outpatient gastroenterology. His symptoms continued to progress and over the following 2 months he had a further ED presentation as well as a second normal CT scan, and he was discharged to the community with no diagnosis identified.

Following concern from his haematologist about the severity of ongoing symptoms, the patient was admitted to Wellington Regional Hospital to facilitate work up. Wide infective, autoimmune, gastrointestinal and haematological panels were unremarkable. Notably, faecal calprotectin was normal at 33µg/g lowering the suspicion for IBD. The patient underwent colonoscopy, gastroendoscopy and CT enterography with findings as follows. Colonoscopy macroscopically found diffuse mild inflammation of the ileum with sparing of the terminal ileum (Figure 4). Histology suggested active mild ileal inflammation. His colon was normal on endoscopic appearance and showed lymphocytic colitis on biopsy. Upper gastrointestinal endoscopy was normal on endoscopic appearance and biopsy. CT enterography was limited by suboptimal bowel distention, but no gross abnormalities were seen despite this.

Given these findings were not typical for Crohn's disease, the differential included gastrointestinal vasculitis, and the patient was briefly started on IV methylprednisolone after his colonoscopy. Vasculitis screening was non-specific: ANA was positive (titre of 1:1320 with homogenous pattern) but the remaining panel including ENA, anti-dsDNA, anti-GBM antibodies and ANCA were normal. Both haematology and gastroenterology noted that the patient's symptoms were disproportionate to that expected in lymphocytic colitis. However, given minimal improvement on IV methylprednisolone he was swapped to oral budesonide, and his omeprazole changed to famotidine due to the possibility of medication-induced lymphocytic colitis. With a subsequent improvement to his pain and bowel motions, the patient was discharged on oral budesonide and analgesics.

Five days after this discharge from Wellington Regional Hospital, the patient re-presented to Wairarapa ED, with the presentation as described above, and passed away. After his death, histology from the resected terminal ileum was reviewed and identified small artery vasculitis in the terminal ileum.

**Figure 2:** Operative specimen photo showing bowel perforations in ischaemic ileum.



**Figure 3:** Transition point of ischaemic—dilated bowel.



Figure 4: Colonoscopy showing mild ileitis.

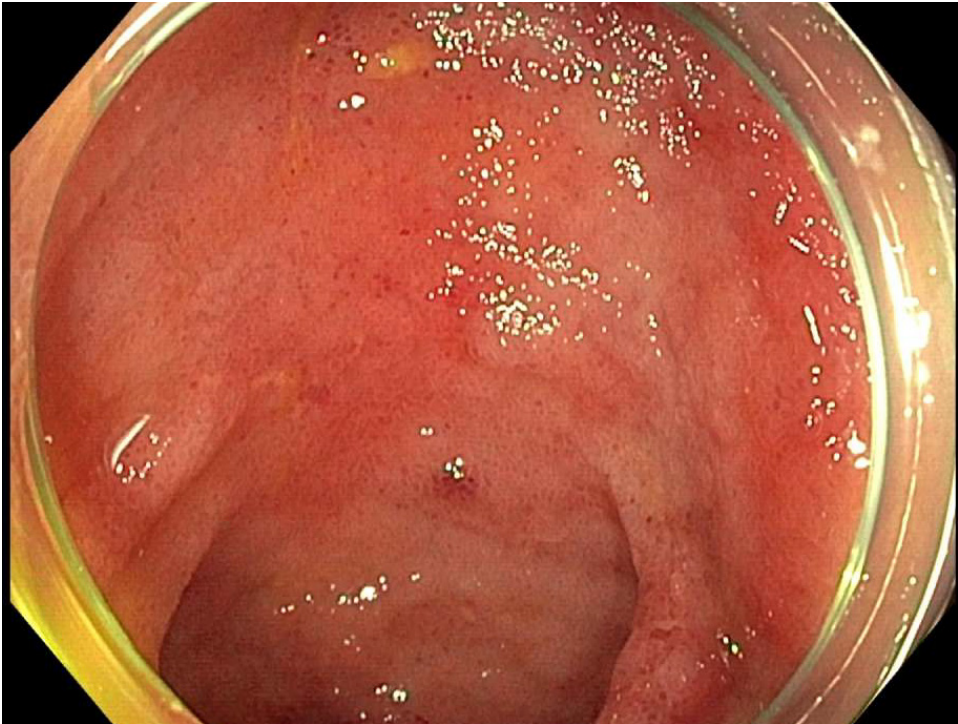
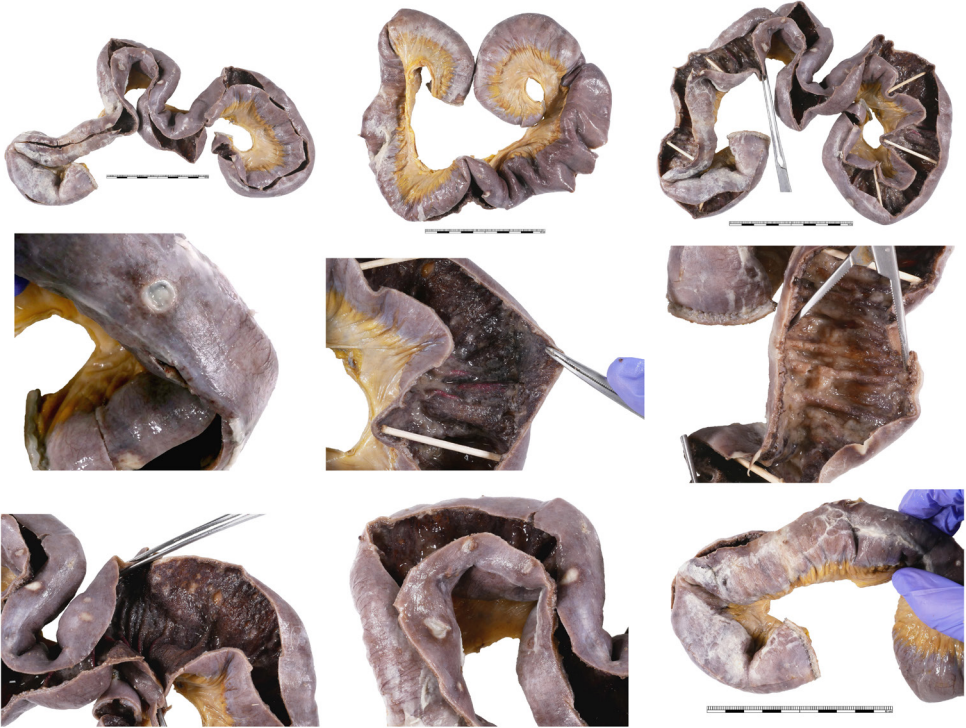


Figure 5: Terminal ileum specimen—showed small artery vasculitis on histology.



## Discussion

Gastrointestinal involvement is common in systemic vasculitic conditions such as Henoch-Schönlein purpura, Polyarteritis nodosa and ANCA-associated vasculitis; however, cases of localised gastrointestinal vasculitis are rare and documented in only a few case reports and small case series. Localised gastrointestinal vasculitis (LVGT) is considered to be extremely rare, a difficult diagnosis to make and is associated with high morbidity and mortality.<sup>1,2</sup>

Salvarani et al. published a series on 18 patients with confirmed LVGT via histology or highly suspicious radiological features between 1996 and 2007 at the Mayo Clinic.<sup>3</sup> The study found abdominal pain as the most common symptom, with 17 of 18 patients complaining of abdominal pain, usually severe. Other common symptoms included nausea, vomiting, diarrhoea, weight loss, melaena, haematochezia and abdominal angina. Salvarani et al. found no specific laboratory findings that were consistently abnormal for their patients. Radiologically, 15 of the 18 patients had undergone some form of abdominal angiography (catheter-based, MRI-based or CT-based), and 14 of these patients had radiological features suggestive of gastrointestinal vasculitis. However,

it must be noted that these features were also used in the inclusion criteria for the study. Of their 18 patients, seven died as a result of their illness during the period of the study; 10 of the 18 patients received medical therapy with immunosuppression and three (30%) of these patients died, whereas four (50%) of those patients who did not receive medical therapy died.<sup>3</sup> Other case series would also suggest LVGT can progress to systemic multi-organ vasculitis. Burke et al. studied 63 patients with LVGT, and during longitudinal follow up, six patients developed systemic vasculitis.<sup>4</sup>

In summary, our patient died from a bowel perforation caused by LVGT of the ileum, confirmed by histology. He had no evidence of other organ involvement in his disease process. LVGT is a rare diagnosis and could only have been confirmed by a full thickness biopsy or indicated towards by abdominal angiography. CT angiography during his acute illness showed only subtle changes to the SMA. His presentation is in keeping with the small case series published on this topic; however, the rarity of this condition and sizes of these case series make it difficult to relate their findings on management and prognosis to this case.

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**COMPETING INTERESTS**

Nil.

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