Pneumatosis intestinalis: a rare manifestation of lupus enteritis

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DESCRIPTION

A 30-year-old female with systemic lupus erythematosus (SLE) presented with high-grade fever, myalgia, diffuse abdominal pain and recurrent vomiting. There was no history of diarrhoea or haematochezia. She was a known case of 'minor organ' SLE with mucocutaneous involvement, leucopaenia and arthritis and had been stable on low-dose steroids, non-steroidal anti-inflammatory drugs and hydroxychloroquine for last 2 years. Laboratory investigations revealed anaemia, leucopaenia, thrombocytopenia, proteinuria, active urinary sediments, antinuclear and anti-dsDNA antibody positivity, transaminitis and a high lactate dehydrogenase. Antiphospholipid antibody profile was negative. Urine culture grew *Klebsiella* and she initially responded to antibiotics. However, she subsequently deteriorated on day 5 of admission, with persistent bilious vomiting, diffuse epigastric pain with tenderness and sluggish bowel sounds, along with mild proximal muscle weakness. Ultrasonography (USG) of abdomen showed bowel wall oedema of 16 mm with loculated air foci (figure 1). CT scan of abdomen revealed dilated, oedematous bowel loops and circumferential collections of air adjacent to the lumen that ran parallel with the wall of the bowel—suggestive of pneumatosis intestinalis (PI) (figure 2).

A diagnosis of lupus enteritis was made and she was treated with pulse methylprednisolone for 3 days, followed by cyclophosphamide. Nutrition was maintained parenterally. Her symptoms started improving and bowel sounds normalised by day 12. Repeat ultrasound showed significant reduction in bowel wall oedema. Oral fluids were reintroduced very gradually, followed by transition to semisolid and then solid diet. She responded to treatment and was discharged.

Lupus enteritis, though infrequent, is a serious complication of SLE. In a large case series of lupus enteritis, it was a presenting manifestation in up to 10% of patients with SLE.¹ It may be associated with vasculitis in other organs and usually indicates an active disease, warranting a swift diagnosis and aggressive immunosuppression. Antiphospholipid antibodies have been associated with increased likelihood of thrombosis, leading to poorer outcomes. Abdominal pain is the most common and cardinal symptom, while nausea, vomiting and diarrhoea with/without fever can also be present. Severe gastrointestinal (GI) bleed, thrombosis and gangrene of intestines are dreaded and almost fatal complications.

Figure 1 USG abdomen showing circumferential hypoechoic thickening (bracket) of bowel wall (B) displaying brightly echogenic foci within suggestive of air loculi pneumatosis (arrows) (A). USG, ultrasonography.

Though CT scan is the investigation of choice, bowel wall oedema on abdominal ultrasound is quite indicative of lupus enteritis. Imaging findings include intestinal wall thickening, dilated bowel loops, abnormal enhancement of the bowel wall (target sign), occlusion in the mesenteric circulation/mesenteric abnormalities (including the comb's sign) and ascites on contrast-enhanced CT (figure 1).²

PI, though not uncommon in clinical practice, is rarely encountered with SLE. The spectrum of presentation varies from asymptomatic (common), to bowel wall perforation, obstruction, GI haemorrhage and death. The investigation of choice is abdominal CT, though PI can infrequently be detected on erect abdominal X-rays and by

Figure 2 (A–D) Transverse and sagittal sections of contrast CT abdomen showing dilated small bowel loop displaying circumferentially thickened and oedematous bowel wall with serosal and mucosal enhancement, giving a target appearance (green arrow) (B, D-yellow arrow). Few loops display intramural air loculi suggestive of pneumatosis (A, C- orange arrows).

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Images in...

ultrasound. Endoscopy and endoscopic ultrasound are considered the gold standard to diagnose PL.³

Lupus enteritis is steroid-responsive and hence high-dose steroids with bowel rest and parenteral nutrition can help avoiding surgery. As our patient had active nephritis, cyclophosphamide according to the modified National Institute of Health (NIH) protocol was given.

Learning points

- ▶ Lupus enteritis should be suspected in all patients with lupus presenting with abdominal pain.
- ▶ Lupus enteritis is usually a harbinger of active lupus, look for vasculitis and other systems involvement.
- ▶ Pneumatosis intestinalis is an uncommon manifestation of lupus enteritis.
- ▶ Ultrasonography is point of care and can help in early diagnosis.

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patient, the CT and USG reporting. KC, RRS and AW were involved in the diagnosis and management of this patient.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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REFERENCES

- 1 Tian X-P, Zhang X. Gastrointestinal involvement in systemic lupus erythematosus: insight into pathogenesis, diagnosis and treatment. *WJG* 2010;16:2971–7.
- 2 Janssens P, Arnaud L, Galicier L, *et al*. Lupus enteritis: from clinical findings to therapeutic management. *Orphanet J Rare Dis* 2013;8:67.
- 3 Ling F, Guo D, Zhu L. Pneumatosis cystoides intestinalis: a case report and literature review. *BMC Gastroenterol* 2019;19:176.

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