Protein-losing enteropathy as a rare manifestation of systemic lupus erythematosus

David Michael Ng ,[1] Kenny Sek,[2] Johannes Nossent[1,3]

[1]Rheumatology, Sir Charles Gairdner Hospital, Nedlands, Western Australia, Australia
[2]Nuclear Medicine, Sir Charles Gairdner Hospital, Nedlands, Western Australia, Australia
[3]Rheumatology Unit, School of Medicine, The University of Western Australia, Perth, Western Australia, Australia

Correspondence to
Dr David Michael Ng;
david.mkc.ng@gmail.com

Accepted 16 August 2023

DESCRIPTION

A woman in her early 20s presented to hospital for investigation of 2 months of worsening dysphagia, abdominal pain, vomiting and 20 kg weight loss. She was diagnosed with systemic lupus erythematosus (SLE) 3 years earlier by a rheumatologist with predominant manifestation of polyarthritis. Her ongoing treatment included daily prednisolone 5 mg and hydroxychloroquine 400 mg and weekly methotrexate 20 mg.

On examination, she was peripherally oedematous and had bilateral pleural effusions confirmed on chest X-ray. There was severe hypoalbuminaemia (23 g/L) with minimal renal loss (microalbuminuria), unremarkable urinary sediment and normal renal and liver function tests (including international normalised ratio). Lipid profile revealed normal total cholesterol (3.0 mmol/L) and mild hypertriglyceridaemia (2.3 mmol/L). She had raised inflammatory markers (erythrocyte sedimentation rate >140 mm/hour), strongly positive dsDNA Ab (6450 IU/mL) and hypocomplementaemia (C4 0.11 g/L, C3 0.59 g/L). Extractable nuclear antigen panel was negative. Recent gastroscopy and manometry were unremarkable and no intra-abdominal pathology was identified on CT.

The patient proceeded to technetium-99m human serum albumin (Tc-99m HSA) scintigraphy which demonstrated small bowel activity consistent with protein-losing enteropathy (PLE; figure 1).

Histopathology from repeat gastroscopy excluded other causes of gastrointestinal disease, with duodenal biopsy negative for Whipple's disease and demonstrating no evidence of active inflammation, villous blunting, intraepithelial lymphocytosis, dysplasia or malignancy. Faecal culture and PCR was negative. Serological testing for HIV, viral hepatitis and coeliac disease was negative, as was tuberculosis interferon gamma release assay testing.

During her admission, the patient experienced a severe multiorgan SLE flare including serositis, polyarthritis, bilateral recurrent laryngeal nerve palsy and anaemia (complicated by *Staphylococcus hominis* empyema and bacteraemia), receiving treatment with intravenous pulse corticosteroids, mycophenolate and rituximab. Infection was managed with drainage and antibiotics and she was supplemented with albumin, electrolyte replacement and furosemide. Her symptoms gradually improved and peripheral oedema resolved. She was discharged home 7 weeks following presentation and at 6-week and 6-month follow-up was in clinical remission.

PLE is a rare manifestation of SLE with the literature consisting of case series and small retrospective studies.[1] The pathogenesis of SLE-related PLE remains unclear but is postulated to include mucosal ulceration, increased mucosal capillary permeability or lymphangiectasia.[2] It is typically characterised by oedema, hypoalbuminaemia and evidence of gastrointestinal protein loss, with independent risk factors reported to include anti-SSA seropositivity, hypoalbuminaemia and hypercholesterolaemia.[2 3] Diagnosis should be confirmed using Tc-99m HSA scintigraphy, which has a high level of diagnostic specificity and can localise sites of protein leakage.[4] Other causes of hypoalbuminaemia (eg, nephrotic syndrome, liver disease, malnutrition) and gastrointestinal conditions leading to villous atrophy (eg, tropical sprue, Crohn's disease, infection) should

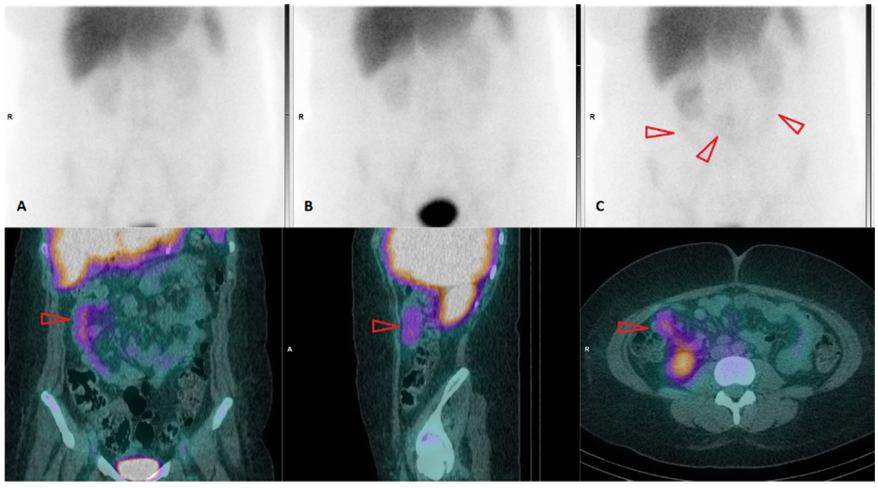

Figure 1 Representative anterior projection planar images of the abdomen at (A) 1 hour, (B) 2 hours and (C) 4 hours, with the 4-hour images showing several regions of faint bowel activity (arrowheads). Fused single-photon emission computed tomography (SPECT)/CT images localises the activity to the small bowel (arrowheads).

To cite: Ng DM, Sek K, Nossent J. *BMJ Case Rep* 2023;**16**:e256680. doi:10.1136/bcr-2023-256680

be excluded. Supportive care and nutritional supplementation are important in the management of PLE; however, definitive treatment involves addressing the underlying SLE.

In our case, the clinical and biochemical findings were consistent with a diagnosis of PLE and confirmed using Tc-99m HSA scintigraphy. Although the patient's duodenal biopsy was normal, the exclusion of other causes and the temporal relationship between her severe SLE flare with the onset and resolution of PLE is suggestive of the underlying cause. The patient remains in clinical remission with no recurrence of PLE since achieving disease control.

Learning points

- Protein-losing enteropathy is a rare gastrointestinal manifestation of systemic lupus erythematosus characterised by oedema, hypoalbuminaemia and evidence of gastrointestinal protein loss.
- Technitium-99m human serum albumin scintigraphy can confirm the diagnosis of protein-losing enteropathy and localise sites of protein loss.
- Definitive treatment requires achieving control of the underlying systemic lupus erythematosus.

Contributors The following authors were responsible for drafting of the text, sourcing and editing of clinical images, investigation results, drawing original diagrams and algorithms, and critical revision for important intellectual content: DMN, KS and JN. The following authors gave final approval of the manuscript: DMN, KS and JN.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Consent obtained directly from patient(s).

Provenance and peer review Not commissioned; externally peer reviewed.

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.

ORCID iD
David Michael Ng http://orcid.org/0000-0001-5436-6380

REFERENCES

1 Frittoli RB, Vivaldo JF, Costallat LTL, *et al*. Gastrointestinal involvement in systemic lupus erythematosus: A systematic review. *J Transl Autoimmun* 2021;4:100106.
2 Peng L, Li Z, Xu D, *et al*. Characteristics and long-term outcomes of patients with lupus-related protein-losing Enteropathy: A retrospective study. *Rheumatol Immunol Res* 2020;1:47–52.
3 Law ST, Ma KM, Li KK. The clinical characteristics of lupus related protein-losing Enteropathy in Hong Kong Chinese population: 10 years of experience from a regional hospital. *Lupus* 2012;21:840–7.
4 Chen Z, Li M-T, Xu D, *et al*. Protein-losing Enteropathy in systemic lupus erythematosus: 12 years experience from a Chinese academic center. *PLoS ONE* 2014;9:e114684.