

## Case Report

# Debilitating chronic diarrhoea with resistant ascites: a case of lupus protein-losing enteropathy

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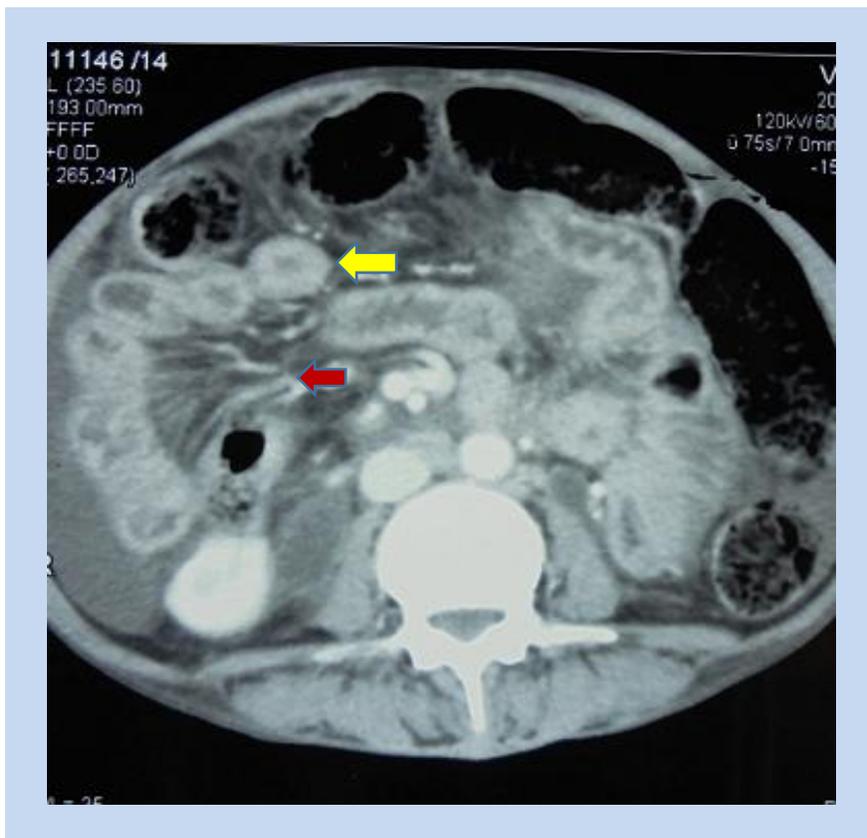
## Introduction

Gastrointestinal manifestations occur in approximately 25 to 40 percent of patients with systemic lupus erythematosus (SLE)<sup>1</sup>. Protein-losing enteropathy (PLE) in SLE, termed lupus protein-losing enteropathy (LUPLE), is a well-recognized clinical entity which is increasingly being noted in a number of case reports both locally and internationally<sup>2,3</sup>. PLE is characterized by excessive loss of serum proteins from the gastrointestinal mucosa which subsequently leads to significant hypoalbuminaemia<sup>1</sup>. Recently it is being frequently identified as the presenting feature of SLE<sup>2,3</sup>. Ascites in SLE commonly occurs as a consequence of its complications such as PLE, nephrotic syndrome, constrictive pericarditis and Budd-Chiari syndrome as well as a part of the disease itself in the forms of acute and chronic lupus peritonitis<sup>6</sup>. We report a rare case of a Sri Lankan female extensively investigated for long term recurrent diarrhoea, vomiting and gradual onset massive ascites who was ultimately diagnosed to have SLE associated PLE and lupus enteritis.

## Case report

A 35 year old female from Avissawella presented to the National Hospital Sri Lanka in early 2013 with a history of one month duration watery diarrhoea, abdominal pain and progressive abdominal distension associated with anorexia and weight loss. On physical examination she was emaciated with evidence of non-scarring alopecia, healed oral ulcers, non-deforming synovitis of interphalangeal joints, pallor and gross ascites without abdominal organomegaly. She had no history of fever, photosensitivity, skin rash or lymphadenopathy. Laboratory work up revealed a haemoglobin concentration of 9.2g/dl, erythrocyte sedimentation rate (ESR) of 94 mm 1<sup>st</sup> hour and hypoalbuminaemia (serum albumin - 22g/L). Ascitic fluid analysis showed an exudate (peritoneal fluid protein - 3.36g/dl) with lymphocytic predominance and serum ascitic albumin gradient (SAAG) <1.1 g/dl suggestive of serositis. Her anti-nuclear antibody (ANA) was positive at a titre of 1:640, anti-double stranded deoxyribonucleic acid (ds-DNA) was positive and serum immunoglobulin E (IgE) level was markedly elevated at 1194 IU/ml. The contrast enhanced computed tomography (CECT) scan of the abdomen revealed thickened small bowel wall >4mm and increased attenuation of omental fat. She underwent double balloon enteroscopy which demonstrated oedematous mucosa with whitish spots reported as more in favour of PLE. Two fragment jejunal biopsy histology was unremarkable and a technetium 99m-labeled albumin study or alpha-1 antitrypsin clearance could not be carried out because of unavailability of the investigation facility. Patient was diagnosed with SLE associated lupus enteritis and PLE and was commenced on prednisolone,

azathioprine and hydroxychloroquine which resulted in dramatic resolution of symptoms and laboratory parameters with reduction of SLE disease activity index from 14 to 5, within two months.



**Figure 1: Contrast-enhanced CT scan showing severe diffuse multifocal bowel wall thickening, demonstrating the target sign (yellow arrow) and increased attenuation "haziness" of omental fat (red arrow), two out of the three cardinal imaging signs of lupus enteritis**

## Discussion

The case is important in that it is an unusual presentation of a relatively common autoimmune disease. Reaching the final diagnosis was delayed because of the absence of the classic signs of SLE as well due to the rarity of the entity. The chronic diarrhoea could probably be attributed to PLE resulting in hypoalbuminaemia and consequent ascites with contributing lupus enteritis causing intestinal capillary leakage and mucosal oedema. Though protein loss through gastrointestinal tract can be confirmed by Tc-99m albumin scintigraphy or stool alpha 1 antitrypsin clearance<sup>4</sup>, these investigations were not available. The diagnosis of SLE was confirmatory and the raised IgE level, endoscopic appearance of the jejunal mucosa and CECT abdomen findings<sup>2,3</sup> along with the striking clinical response to immunosuppression, strongly supported the final diagnosis of LUPLE.

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