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CASE REPORT

Outcome of surgical resection for protein-losing enteropathy in systemic lupus erythematosus

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Abstract Protein-losing enteropathy (PLE) is an uncommon manifestation associated with systemic lupus erythematosus (SLE). Here, a case with SLE and concomitant hypoalbuminemia is reported. Technetium-99m albumin scintigraphy demonstrated a localized lesion in the ascending colon, and the diagnosis of SLE-related PLE was established. Due to a poor response to medical treatment, this patient received surgical resection, but relapse still developed later on. Recurrent protein-lose from the remaining of the colon was documented by repeated images. This report discusses the management of SLE-related PLE and the role of nuclear medicine scintigraphy in the investigation of PLE.

Keywords Albumin · Protein-losing enteropathy · Systemic lupus erythematosus · Tc-99m

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Introduction

Protein-losing enteropathy (PLE) is characterized by the loss of serum protein from the gastrointestinal tract, and is usually associated with disease activity in other organs. Systemic lupus erythematosus (SLE) is a multi-system disease, and protein-losing enteropathy is an uncommon manifestation of SLE [1, 2]. Various treatments of SLE-related PLE have been reported, but most of it relies on subjective observation. Management with corticosteroids alone or combined with immunosuppressive treatment remains the most commonly reported method of treatment [1, 3, 4]. However, the most favorable therapy is still unidentified due to the lack of controlled clinical trials.

We report herein a 27-year-old female with previously diagnosed SLE, who presented with PLE. Her clinical manifestations persisted after medical management with corticosteroids. The Technetium-99m human serum albumin (Tc-99m HSA) scintigraphy showed a localized area of protein loss to the ascending colon. Following surgical resection effectively relieved her symptoms. However, recurrent symptoms and signs developed again 6 months later, and repeat nuclear medicine studies demonstrated protein-losing originating from the remaining colon.

There is no published literature reported previously on the subject of surgical intervention for SLE-related PLE.

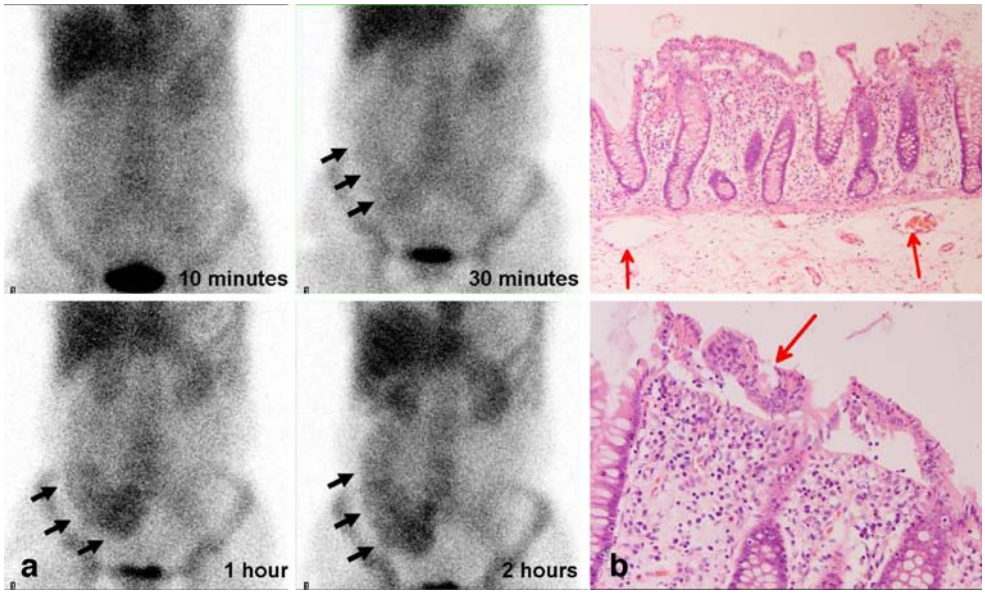

Fig. 1 **a** Tc-99m HSA scintigraphy of abdomen. An area of dilated bowel with radiotracer accumulation to the right lower quarter of abdomen (*arrows*) is revealed. This demonstrates a localized segment of protein loss to the ascending colon. No evidence of involvement to the rest of the colon was found. **b** Pathology photographs of the resected colon. *Upper* Atrophic mucosa, edematous change to the lamina propria and submucosa were present. Some dilated capillaries and lymphatic duct were found (*arrows*). *Lower* Detached columnar epithelium, thickened basal lamina, capillary proliferation and some plasma cells and lymphocytes infiltration in lamina propria were identified (*arrow*). These findings are consistent with the clinical manifestation of PLE

Until now, limited reports regarding Tc-99m HSA scintigraphy in the SLE-related PLE had been documented [1, 5, 6]. We now report on our case and review the literatures.

Case report

This 27-year-old female SLE patient had been diagnosed and managed via medical therapy for 4 years. Recently, hypoalbuminemia developed with the clinical manifestation of acute abdominal pain, abdominal distention, massive ascites, and pleural effusion. Laboratory studies revealed a severely diminished serum albumin level to 1.1 gm/dL (normal range 3.9–5.1 gm/dL) and decreased total protein to 4.7 gm/dL (normal range, 6.4–8.1 gm/dL); the others such as blood routine studies and serum electrolytes were essentially within normal range. Urinalysis confirmed that there was no urinary excretion of protein found. In addition, malnutrition and hepatic problems were also excluded from history, physical examination, and blood examination. Tc-99m HSA scintigraphy was performed and protein leakage from the ascending colon was demonstrated (Fig. 1a). Under the impression of hypoalbuminemia caused by PLE, medical treatment with hydroxychloroquine sulfate 200 mg/day and methylpredisolone 4 mg/day was administrated. However, her symptoms persisted.

Due to poor response to medical therapy and image evidence of localized protein-losing area to the proximal

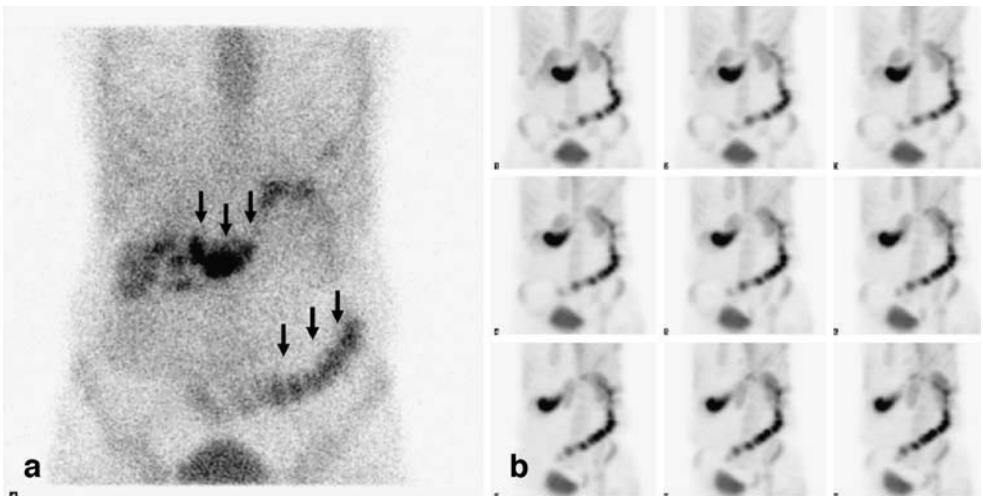

Fig. 2 Six months after surgical resection of the ascending colon, Tc-99m HSA scintigraphy was performed again for the re-evaluation of the relapsed symptoms. This image result clearly demonstrates protein loss from the rest of the colon, especially to the transverse colon. **a** Planar images, **b** 3D reconstructed SPECT images. Areas with radiotracer retention are shown (*arrows*)

portion of ascending colon, surgical resection was arranged consequently. The pathology report proved that this is consistent with the PLE (Fig. 1b).

After surgical resection, her general condition improved. The clinical complaints resolved, and the serum albumin level achieved the level of 3.0 gm/dL and total protein 6.7 gm/dL, respectively. She was discharged with regular follow-up at our outpatient department. Nevertheless, about 6 months after that operation, recurrent abdominal colic manifested again. She was readmitted to the hospital, where all of the symptoms of hypoalbuminemia emerged again. Serum investigations revealed that level of albumin had declined to 1.2 gm/dL and total protein to 2.8 gm/dL. Recheck with Tc-99m HSA scintigraphy demonstrated major leakage from the transverse colon and passage to the descending colon (Fig. 2).

Discussion

Hypoalbuminemia is defined as a diminished serum albumin level. Causes of hypoalbuminemia in a SLE patient could be divided into two major categories: (1) decreased protein synthesis, this could be attributed to deficient protein intake or liver diseases or (2) excessive protein loss, generally followed by nephritis or, less frequently, due to PLE. Further verification can be made by identifying protein loss through the gastrointestinal tract, using methods such as fecal alpha-1 antitrypsin clearance [7], chromium-51 (Cr-51) albumin, or iodine-131 (I-131) polyvinyl pyrrolidone studies. Alpha-1 antitrypsin is a serum protease which is not digested in the gastrointestinal tract it is excreted mostly in its intact form in stools. The 24-h stool clearance of alpha-1 antitrypsin is considered a good indirect measurement of gastrointestinal tract albumin loss. In addition to the indirect evidence for gastrointestinal protein loss, nuclear medicine scintigraphy using the radiolabeled albumin provides a direct, physiological demonstration of bowel leakage. Tc-99m HSA method appears to have good stability, and is appropriate for the clinical application in current practice. Abdominal scintigraphy after intravenous administration of Tc-99m HSA provides visualization of the site of intestinal albumin loss [8]. This presents a useful and stable approach, not only to diagnose of PLE, but also to detect and localize the involved region with PLE [9].

PLE may occur in SLE patients of any age, especially in young female patients. The usual presenting symptoms are pronounced edema and diarrhea; and, in about two thirds of patients, these comprise the initial symptom of SLE [10, 11]. High-dose corticosteroids, with or without an adjunct agent, such as cyclophosphamide (CTX), azathioprine (AZA), or methotrexate (MTX), are often effective in controlling the symptoms [4, 12, 13]. Most cases experience a good outcome after medical management, with relapses occurs in approximately 20–30% of patients [10]. However, our patient did not respond to the corticosteroid therapy. Since the localized protein-losing segment to the ascending colon was presented on the scintigraphic image, surgical resection was performed and it indeed showed sufficient response initially. Unfortunately, her symptoms resumed only 6 months later. Our case showed poor compliance after surgical intervention.

Surgical resection, as a therapeutic approach for PLE in various causes, has been documented with good prognosis [14, 15]. However, no previous report mentioned about this procedure as performed under the condition of SLE-related PLE. Since SLE is a disease classically associated with multisystemic involvement, the mechanisms of protein loss mentioned above are further complicated. Mere resection of the initially involved bowel segment could not eradicate the scourge of complications, and recurrence could be expected.

In conclusion, this report comes out with two major components. First, although surgical resection has been demonstrated to be effective in the management of patients with PLE, it has not been shown to work for SLE-related PLE, and our patient only achieved short-term benefits as well. Secondly, Tc-99m HSA scintigraphy is practical and effective in the detection and localization of PLE. It not only can provide direct evidence of protein loss, it also can identify the involved area. This procedure is easy for a nuclear medicine laboratory to proceed and it can be applied in the initial diagnosis, follow-up evaluation, and relapse check-up of PLE.

Disclosures None.

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