Protein-losing enteropathy caused by spontaneous superior mesenteric artery dissection with thrombosis

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To the Editor,

Protein-losing enteropathy (PLE) is a rare clinical condition caused by continuous protein leakage into the digestive tract, resulting in hypoproteinemia, which can be complicated by edema, ascites, and malnutrition (1). It is observed in association with various disorders. However, its association with isolated spontaneous dissection of the superior mesenteric artery with thrombosis (DSMAT) has not been reported before. We describe a patient who exhibited PLE due to DSMAT, and discuss the possible mechanisms and treatment of refractory hypoproteinemia in this condition.

A previously healthy 51-year-old Chinese man presented to the emergency department (ED) of our Hospital with a 2-hour history of periumbilical pain associated with nausea and vomiting in September 2008. His blood pressure was normal (110/70 mmHg). No tenderness and rebound tenderness was noted. The abdomen had normal bowel sounds on auscultation. Vascular examination revealed normal pulses in the carotid, radial, femoral and pedal arteries. He had a white blood cell count of 7.3×10^9/L ($4.0\text{-}10.0 \times 10^9$/L) and an elevated amylase value of 208 U/L (20-120 U/L). Prothrombin time and partial thromboplastin time were both normal. An abdominal plain X-ray film revealed a nonspecific ileus and an abdominal ultrasound revealed no abnormal findings. He was fasted and was given antibiotics and fluid infusion. He continued to have severe abdominal pain. For suspicion of ischemic enteritis he underwent an enhanced CT scan revealing DSMAT (Fig. 1). Because there were no complications, the patient was treated conservatively with anticoagulation, antibiotics and a nutritional support (2), resulting in partial recovery. Enteral nutritional support using a semi-elemental formula and additional protein supplementation was begun on the 32th day. Unfortunately, despite modifying his diet to include a protein intake of at least 2 g/kg/day, his albumin and protein levels remained low, around 17-24 g/L and 35-55 g/L respectively. Capsule endoscopy revealed small intestinal erosions and stricture. The capsule became impacted at this point. After adequate preparation the patient was taken to the operating room where an ileocecectomy was performed, including removal of 150 cm of ileum. Pathology revealed that both the ileum and the mesentery were whitened and thickened. Microscopical examination disclosed mucosal necrosis, fibrosis and full-thickness inflammatory cell infiltration. In the mesentery, some arterial branches exhibited organized thrombi with recanalization. One week after surgery, his peripheral edema had resolved and his albumin had normalized. The patient had an uneventful postoperative course and was discharged on the 37th post-operative day. At 6-month follow-up, the patient continues to be free of abdominal pain, and gains weight without treatment.

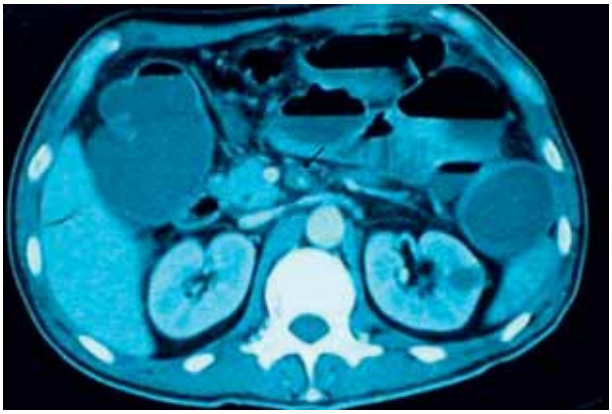

Fig.1. — Angio-CT of the superior mesenteric artery. The intima separates the true and false lumen in the SMA (arrow). Bowel loop dilatation, suggesting ischemic change in the small bowel.

PLE is a severe intestinal loss of proteins leading to a clinical picture characterized by hypoproteinemia, hypoalbuminemia and hypogammaglobulinemia (3). Causes of protein loss from the intestinal tract can be divided into three categories : 1. loss from intestinal lymphatics, which can be primary (primary intestinal lymphangiectasia) or secondary due to lymphatic obstruction (as seen in retroperitoneal fibrosis and lymphoma) or increased lymphatic pressure (as seen in constrictive pericarditis and portal hypertension) ; 2. loss from the mucosal surface after disruption of the mucosa by infectious, immunological, or vasculitic disorders ;

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and 3. loss for metabolic reasons due to congenital disorders of glycosylation or heparan sulfate deficiency (4). We believe that the mucosal necrosis and fibrosis due to DSMAT are responsible for the protein loss in the present case. It was confirmed by capsule endoscopy and histological examination, which demonstrated evidence of disruption of the mucosa. Furthermore, the patient underwent ileocecectomy with complete resolution of PLE. These results suggest that DSMAT is the underlying cause of PLE in our patient.

The manifestations of PLE are highly variable because of the different underlying causes. The most important laboratory findings are reduced serum concentrations of albumin, protein and globulins. Although the hypoalbuminemia contributes to the primary clinical manifestation of edema due to diminished plasma oncotic pressure, the reductions of other serum proteins rarely cause clinically significant problems. Our patient initially developed abdominal pain without any classic manifestations of PLE. Hypoproteinemia, hypoalbuminemia and hypogammaglobulinemia developed with the development of disruption of the mucosa, and he failed medical and nutritional treatment. These findings strongly supported the diagnosis of PLE.

Treatment options may include medical, nutritional and surgical intervention. In most cases, medical treatment can only provide temporary improvement in symptoms, and so surgical treatment is the only therapeutic option. However, hypoalbuminemia and decreased immunity increase the morbidity and mortality of operation. Therefore, preoperative and postoperative nutrition support plays an important role in the success of surgery. Normal protein requirements are generally 0.6-0.8 g/kg/day. In PLE, this value may increase to 2.0-3.0 g/kg/day to achieve positive protein balance (1). Although it is always preferable to use the gut for feeding, in certain conditions such as severe erosive disease or intestinal dysmotility, parenteral nutrition support is necessary. In the present case, the use of intermittent intravenous infusions of albumin is unhelpful, then surgery was performed which resulted in resolution of hypoalbuminemia. The results suggest that treatment of PLE targets not only clinical symptoms but also the underlying causes.

To our knowledge, this is the first case of PLE caused by DSMAT. It should be considered as a possible cause of PLE. Surgical resection of disruptive focus seems to be the most effective method of therapy for such patients.

References

1. UMAR S.B., DIBAISE J.K. Protein-Losing Enteropathy : Case Illustrations and Clinical Review. *Am. J. Gastroenterol*., 2009, **104** : 1-7.
2. AMABILE P., OUAÏSSI M., COHEN S., PIQUET P. Conservative treatment of spontaneous and isolated dissection of mesenteric arteries. *Ann. Vasc. Surg.*, 2009, **23** : 738-744.
3. SORIANO J.R., GRIMA E.Z., VIVES M.A., SÁEZ A.O., DOLZ L.M., BONET L.A., PÉREZ M.A., SANZ A.S. Reversal of protein-losing enteropathy after heart transplantation in young patients. *Rev. Esp. Cardiol*., 2009, **62** : 937-940.
4. ALKHOURI N., CARTER-KENT C., MAYACY S., HUPERTZ V., EGHTESAD B., QUINTINI C., FUNG J., RADHAKRISHNAN K. Reversal of protein-losing enteropathy after liver transplantation in a child with idiopathic familial neonatal hepatitis. *Liver Transpl*., 2009, **15** : 1894-1896.