

Remission of protein-losing enteropathy after surgical repair of a giant umbilical hernia in a dialysis patient

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ABSTRACT

We present a 57-year-old male chronic haemodialysis patient with a giant umbilical hernia. After two months on dialysis, incarceration of the hernia with partial intestinal obstruction occurred, and this led to severe hypoproteinaemia secondary to protein-losing enteropathy, caused by increased lymphatic pressure. These problems were resolved following surgical repair of the hernia. There are few such cases in the literature and none involving a haemodialysis patient.

Key-Words:

Haemodialysis; hypoalbuminaemia; protein-losing enteropathy; umbilical hernia.

CASE REPORT

We present a 57-year-old male chronic haemodialysis patient with a giant umbilical hernia. He had active metabolic syndrome with obesity (BMI 36 kg/m²), long-standing high blood pressure, hypertensive heart disease, small-vessel encephalopathy and moderately severe COPD. He was a former cigarette smoker (180 pack/years) and a former drinker of alcohol (40 g/day).

He had undergone two previous umbilical hernioplasties followed by recurrence of the hernia ten

years previously. His chronic renal failure was the result of hypertensive nephroangiosclerosis. At the time of starting haemodialysis, his serum total protein and serum albumin levels were 5.8 g/dL and 2.7 g/dL respectively, and rose to about 7.0 g/dL and 3.1 g/dL respectively, after removal of fluid to improve oedema over the first month of dialysis (Fig. 1).

At month 2, the patient complained of pain from his large umbilical hernia; he had previously been pain-free for many years though he did have a 20-year history of occasional diarrhoea. The size of the hernia had gradually increased. At this time the patient's

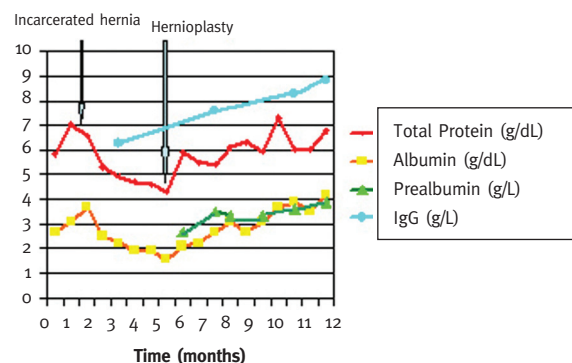


Figure 1
Serum analytical evolution

serum total protein and serum albumin levels were 6.6 g/dl and 3.7 g/dL respectively.

At month 3, the patient developed sensory ataxia as the result of dorsal cord syndrome, and was found to have marked hypoproteinaemia (4.3 g/dL), hypoalbuminaemia (1.6 g/dL) and hypogammaglobulinaemia. Adjusted serum calcium, liver function tests, thyroid function tests, haemoglobin, leukocytes, platelets, vitamin B12, folic acid and coagulation screen were normal. He continued to pass 500 mL of urine per day and total urine protein was 440 mg/24h. His appetite remained good, he was eating well, and was given oral nutritional supplements which were well tolerated.

Mantoux and Quantiferon® (interferon- γ release assay) tests were negative for mycobacteria; serum angiotensin converting enzyme and cortisol were normal; tumour markers were normal; antinuclear antibodies, anti-DNA, anti-neutrophil cytoplasmic antibodies, and extractable nuclear antigen tests were negative; serum complement screen was normal; serum protein electrophoresis showed no monoclonal component, decreased albumin and gammaglobulins, with IgG of 624 mg/dL, normal IgA and IgM, and

increased alpha-1, alpha-2 and beta-globulins. An abdominal ultrasound ruled out portal hypertension. CT and MR scans of brain showed small-vessel encephalopathy, and whole-body CT showed sub-acute intestinal obstruction with distension of the bowel loops due to the large incarcerated umbilical hernia (Fig. 2).

Screening tests for malabsorption were performed. Faecal fat was 7 mg fat/g stools (reference value <65); testing for chymotrypsin in faeces was positive and alpha-1 antitrypsin in faeces was elevated at 1.88 mg/g (reference value <0.3). Other findings were serum IgA anti-transglutaminase antibodies negative; normal serum levels of vitamin B1, B12, folates and copper; normal plasma aluminium; severe serum B6 hypovitaminosis (<3 μ g/L (reference range 4-18). Lumbar puncture was also performed, returning normal blood biochemistry, cell count, antibodies and cultures. Treatment was started with high doses of oral vitamin B6.

At month 4, hernia repair (lateral internal sphincterotomy) was performed. Examination of the hernial sac showed adipose tissue and fibrosis, with no signs of chronic inflammation. Thereafter, the patient's

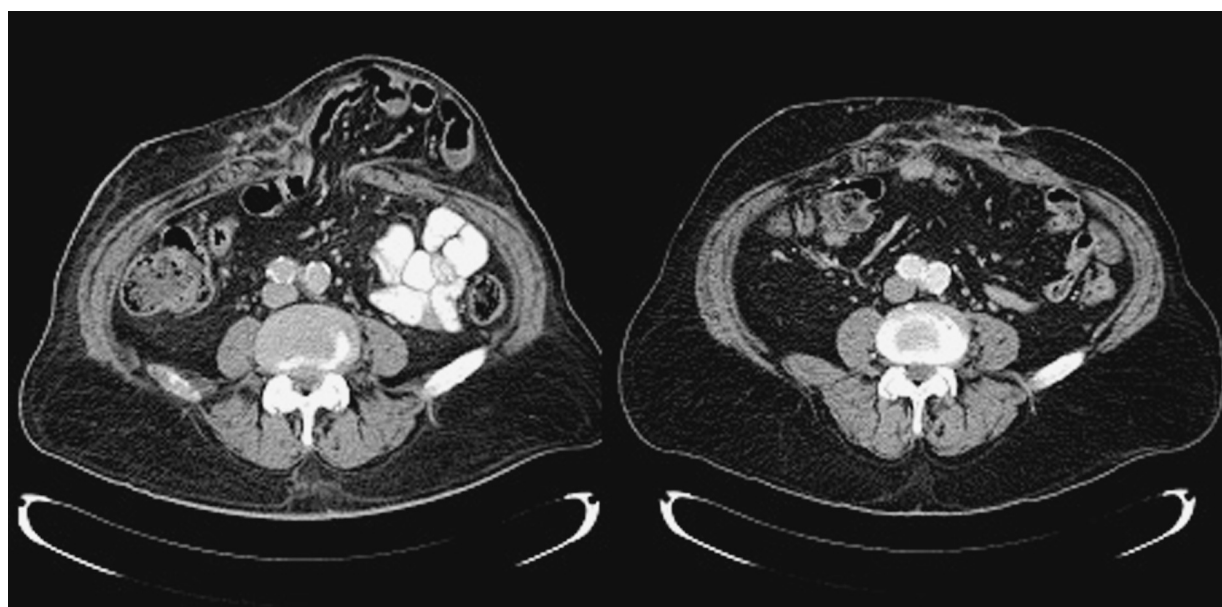


Figure 2

Abdominal CT before hernioplasty (left) and after hernioplasty (right)

motor stability progressively and consistently recovered and his nutritional state improved, with serum total protein rising to 5.9 g/dL and serum albumin to 2.1 g/dL.

Repeat testing performed at month 5 (1 month after surgery), showed that faecal fat, chymotrypsin and alpha-1 antitrypsin had returned to normal. Upper gastrointestinal and small bowel X-ray series showed only non-specific oesophagitis and non-specific changes in duodenum, jejunum and ileum folds.

At month 9, patient had normal intestinal rhythm with no abdominal pain, and gait had returned to normal. Nutritional values were stable (total protein and albumin levels: 6.3 g/dL and 2.7 g/dL respectively), with a progressive increase in IgG to 825 mg/dL and stable haemoglobin and red cell distribution width. Upper and lower GI endoscopy were performed, showing peptidic acid enteropathy and non-specific terminal ileitis seen at colonoscopy. Loop stenosis or villous atrophy were not observed. The abdominal CT scan was repeated, showing no signs of intestinal obstruction or free intra-abdominal fluid. The patient is presently free of digestive symptoms, twelve months after surgery, with normal total serum protein and albumin levels (7.0 g/dL and 4.1 g/dL respectively).

DISCUSSION

This case, a complex one because of co-morbidity and a slowly-evolving disease process, shows the difficulties involved in reaching an accurate diagnosis in some kidney patients. In this man, the combination of hypogammaglobulinaemia and hypoalbuminaemia caused us to suspect protein-losing enteropathy.

The patient was eating well and did not complain of disorders in intestinal rhythm until interrogated directly, which he then attributed to polypharmacy. Neither increased protein needs, due for example to hyperthyroidism, nor low protein synthesis rates due to liver disease or chronic infection, were detected. Initial normoalbuminaemia ruled out hereditary analbuminaemia; there were no findings to justify a diagnosis of hypercatabolism (normal bone series,

no evidence of intestinal lymphoma, infection or trauma), and there was no evidence of a dilutional state (eg SIADH, psychogenic diabetes insipidus). We therefore considered increased protein loss as the most likely cause of hypoproteinaemia.

Having ruled out nephrotic syndrome, burns, bleeding and ascites, the only remaining possible cause of protein loss was a protein-losing enteropathy, presumed to be the result of intestinal lymphangiectasia secondary to the massive umbilical hernia¹; this was confirmed by resolution of the problem following hernia repair. In this condition, increased lymphatic pressure causes a loss of proteins from the epithelial surface of the intestine. The nature of the loss does not depend on the molecular weight of the protein, unlike the glomerular mechanism. The symptoms are loss of iron, immunoglobulins, transport proteins, water-soluble vitamins, trace elements and lipids, leading to inadequate absorption of fats and proteins, and vitamin deficits. The causes of secondary intestinal lymphangiectasia² include constrictive pericarditis, inflammatory intestinal disease, infections, after-effects of chemotherapy and toxins, portal hypertension, Whipple's disease, celiac disease and intestinal lymphoma. In the literature we have found only two similar cases, and neither was a patient on haemodialysis^{3,4}.

Conflict of interest statement. None declared.

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