Case Report

Dialysis-related amyloidosis (DRA) in a patient on CAPD presenting as haemoperitoneum with colon perforation

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Introduction

Dialysis-related amyloidosis (DRA) is a serious complication affecting patients with end stage renal disease and commonly seen in patients undergoing dialysis for an extended period [1]. It also occurs in patients on continuous ambulatory peritoneal dialysis (CAPD). β₂-microglobulin (β₂M) has been implicated as the principal protein component of DRA [2]. Carpal tunnel syndrome is the most common manifestation of DRA, but arthralgia, chronic synovitis, destructive spondyloarthropathy, and pathological fractures of the long bones are frequent [3]. Several cases with visceral involvement have been reported [4]. In this paper, we report a case of DRA presenting as haemoperitoneum with colon perforation.

Case report

A 63-year-old female patient was admitted to hospital because of dyspepsia, anorexia, and malnutrition. She had suffered from diabetic nephropathy since 1988 and had received haemodialysis therapy from May 1992 to November 1995. The dialysis modality was changed to CAPD because of insufficient blood flow of the arteriovenous fistula. Five months prior to admission, she suffered from progressive back pain and weakness of the lower extremities and became unable to walk by herself. Recently, anorexia, dyspepsia and general weakness were aggravated and she was admitted to the hospital.

On admission, she was pale and dehydrated. Her blood pressure was 130/90 mmHg, heart rate 70 beats/min and a body temperature of 37.2°C. Physical examination revealed mild abdominal distension and decreased bowel sound without any peritoneal irritation sign. The muscle strength of the lower extremities was reduced to grade I or II and the muscle mass was atrophied. The deep tendon reflexes were normal. Other findings were unremarkable. Laboratory tests on admission showed urea nitrogen 13.5 mM, creatinine 380.1 pM, calcium 2.1 mM, phosphorus 0.61 mM, total protein 44 g/l, and albumin 15 g/l. The β₂-microglobulin was 380.1 pM, calcium 2.1 mM, phosphorus 0.61 mM, total protein 44 g/l, and albumin 15 g/l. The serum electrophoresis did not have an M peak. Her haemoglobin count was 8.6 g/dl, white blood cell count 11.0 × 10⁹/l, and platelet count 332 × 10⁹/l. Conservative treatment was started.

On the third day after admission, haematemesis was developed and she complained of severe abdominal pain. The CAPD effluent became bloody with a red blood cell count of 0.6 × 10⁹/l and a leukocyte count of 9.0 × 10⁹/l with neutrophilia. Body temperature was 38°C and there was abdominal tenderness with peritoneal irritation signs. Emergency gastrofibroscopy showed multiple shallow bleeding ulceraions throughout the stomach and duodenum. Thoracic radiography revealed a large amount of free air under the diaphragm.

With the clinical diagnosis of upper gastrointestinal bleeding with viscous perforation, emergency explorolaparotomy was performed. The peritoneal cavity contained ~500 ml of seropurulent exudate. The entire wall of the small bowel and colon were thickened. The perforation was in the hepatic flexure of the transverse colon. The perforation was 2.0 cm in diameter and had a very thin and friable margin. Right hemicolectomy was conducted. The anterior wall of the body of the stomach was opened. There was active bleeding from the base of the ulcer on the posterior wall of the antrum. Suture ligation was done and gastrotomy was closed.

The patient’s post-operative course was downhill.
and she died on the 8th day after the surgery from septic shock and multiple organ failure.

On gross examination of the resected colon, the bowel wall was thickened and multiple shallow ulcerations ranging from 2 to 50 mm were noted (Figure 1). On staining with haemotoxylin and eosin (H&E), amorphous, eosinophilic material was deposited in the submucosa and perivascular area of the intestine (Figure 2). The deposits were positive for Congo Red staining and had apple-green birefringence under polarizing microscopy (Figure 3). Immunohistochemical study with anti-\(\beta_2\)-M (Biomeda Corporation, Foster City, CA, USA) and was positive (Figure 4).

**Discussion**

DRA was first reported in 1980. Accumulation of \(\beta_2\)M, a polypeptide with a molecular weight of 118 kDa which is normally present on the surface of class I major histocompatibility antigens, is most likely responsible for this disorder [5]. In general, DRA develops in patients undergoing long-term haemodialysis. Although DRA has been reported in a case of chronic renal failure who had never been dialysed, it usually develops in patients who have received dialysis therapy for >5 years [1].

These amyloid fibrils have a strong predilection to accumulate in tendons, periarticular structures, at the end of long bones, and within the vertebral disks. The most common clinical manifestation is carpal tunnel syndrome and other musculoskeletal abnormalities are frequent complaints in patients with DRA. Recently, several studies on the distribution of dialysis related amyloid deposits have shown that there is extensive involvement of viscera such as heart, lung, liver, kidney, lymph node, stomach and intestine [6]. There are few reports of gastrointestinal involvement of DRA. And it has been suggested that DRA can cause serious gastrointestinal complications such as haemorrhage, intestinal perforation, obstruction, and infarction [7–10].

Our case was a very unusual one. Although the patient complained of back pain and weakness of lower extremities, there was no radiological evidence of amyloidosis on high resolution skeletal roentgenogram. The duration of dialysis therapy was \(~3\) years. According to other reports, the symptoms and signs of DRA appear in patients with long-term dialysis of at least 5 years. The duration of dialysis was not long enough to suspect DRA.
In conclusion, dialysis related amyloid can be deposited in solid organs and manifest itself in various ways. DRA should be considered in patients receiving haemodialysis or on CAPD who have unexplained gastrointestinal symptoms. In addition, DRA may develop early in the course of dialysis.

References


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