CASE REPORT

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Steroid Responsive Abdominal Cocoon Syndrome Secondary To Peritoneal Dialysis; A Rare Case Presentation

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Abstract

Abdominal cocoon syndrome is a rare condition that defines the total or partial encapsulation of the small bowel loops by a fibcrocollagenous membrane, primarily or secondarily, with inflammatory infiltrate that leads to acute or chronic obstruction. While this condition may become manifest in an idiopathic-primary form, it can also occur secondary to interventions such as abdominal surgery, chronic peritoneal dialysis, liver transplantation, ventriculoperitoneal shunt or medications.
We aimed to present a case diagnosed with abdominal cocoon syndrome induced by peritoneal dialysis, who showed a good clinical response to steroid treatment.

Keywords: Abdominal cocoon Syndrome, Peritoneal Dialysis, Steroid Treatment

Introduction

Sclerosing encapsulated peritonitis, a rare cause of acute abdomen or intestinal obstruction, was first described in 1868 and the definition of peritonitis chronica fibrosa incapsulate was used (1). Abdominal cocoon syndrome was first systematically described in Foo et al. made by, in 1978 (2). Factors affecting the clinical signs of ACS are duration and severity of the disease, underlying causes and immunological condition of patient. ACS is most commonly characterised-by recurrent intestinal obstruction attacks (3).

Case Report

A sixty-three-year-old female patient who presented to the emergency service with the complaints of abdominal pain, nausea, vomiting and constipation was admitted to the General Surgery clinic for a provisional diagnosis of ileus due to the presence of air-fluid levels on her erect plain abdominal radiograph (Figure-1). The patient was operated since her clinical picture did not improve after 2 days of medical follow-up. The operation was terminated after a peritoneal biopsy was taken due to the intraoperative finding of diffuse peritoneal adhesions, the patient was consulted to our department and transferred to the nephrology service.

The patient, who reported receiving peritoneal dialysis for 12 years and experiencing several peritonitis episodes in the last 2-3 years in her history, was frequently hospitalized for this reason. The patient, whose peritoneal fluid cultures did not demonstrate bacterial growth, was empirically started on vancomycin. She was most recently hospitalized 2 months ago due to peritonitis and was switched to a hemodialysis program after the removal of her peritoneal catheter. The patient, whose abdominal pain persisted, showed diffuse tenderness and guarding on abdominal physical examination. Blood tests were as follows WBC: 12200 10^3/uL, creatinine: 2.7 mg/dl, BUN: 18mg/dl, Na: 136 mmol/l, K: 3 mmol/l, P: 2.5 mg/dl, Ca: 8.1 mg/dl, CRP: 166 mg/L.

On ultrasonography (USG), hypo-echoic areas measuring 3 cm in maximal diameter were observed between the anterior abdominal wall and the parietal peritoneum. Computerized tomography (CT) revealed a septated fluid collection in the anterior abdominal wall that showed subcutaneous extension and measured 3 cm in maximal diameter, and air-fluid levels in ileal loops in the left lower and middle abdominal quadrants (Figure-2). The performed USG

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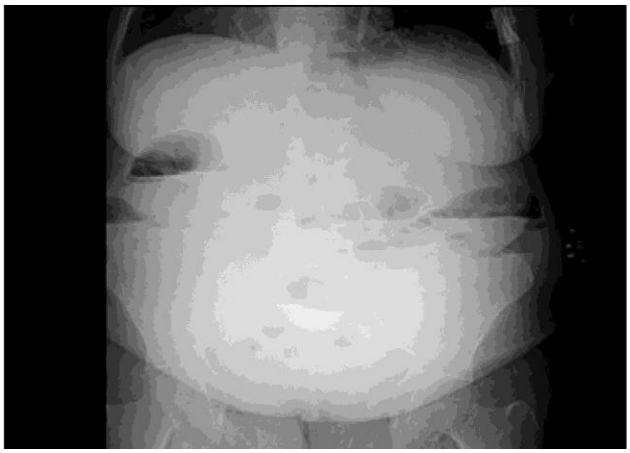

Fig 1. Air-Fluid Levels on Erect Plain Abdominal Radiograph

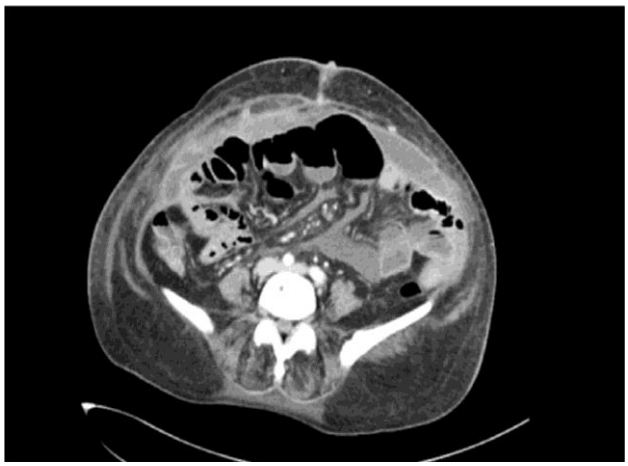

Fig 2. CT Showing Septated Fluid Collection İn The Anterior Abdominal Wall

and abdominal CT were consistent with abdominal cocoon syndrome.

In the peritoneal biopsy of the patient, there was fibroadipose-vascular tissue with a marked increase in fibrotic tissue. Considering the findings of the biopsy indicating the occurrence of an infectious and fibrotic process alongside the clinical and radiologic findings, the patient was diagnosed with abdominal cocoon syndrome.

The patient was commenced on 1 mg/kg (60 mg) methylprednisolone. Her abdominal pain started subsiding on the 7th day of the treatment. Oral intake resumed. Defence and guarding were not detected on examination. Following the administration of 1 mg/kg methylprednisolone for 3 weeks, the dose was tapered by 4 mg per week and the patient was discharged by planning a minimum of 6 months-1year of maintenance therapy on a dose of 8 mg. The erect plain abdominal radiograph of the patient who attended a follow-up examination 20 days later and did not have any complaints, was unremarkable. The methylprednisolone regimen was continued as

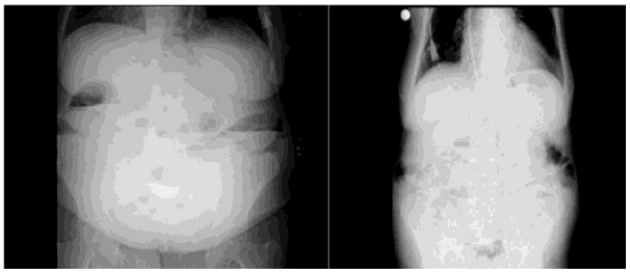

Fig 3. Patient's Erect Plain Abdominal Radiographs At Admission and Post-Discharge

planned. The patient's laboratory parameters (Table 1) at admission, on the 10th day of treatment and at 3 months post-discharge, and erect plain abdominal radiographs at admission and post-discharge (Figure 3) were compared.

Discussion

The mechanism underlying abdominal cocoon syndrome has not yet been completely revealed. It is thought to occur due to peritoneal capillary angiogenesis with the proliferation and hyperplasia of peritoneal mesothelial cells resulting from inflammation in the peritoneum (4).

Abdominal cocoon syndrome is rarely encountered. Its prevalence varies between 0.7 and 13.6 in 1000 among patients receiving long-term peritoneal dialysis (5,6).

The primary risk factor is receiving long-term peritoneal dialysis (5). Experiencing severe peritonitis episodes constitutes another risk factor. The composition of the dialysate fluid also pose risk. Particularly, high glucose and acetate contents are thought to increase the likeliness of the occurrence of abdominal cocoon syndrome. Among medications, the use of beta blockers and calcineurin inhibitors is also considered as risk factors (7). In our case, our patient had been receiving peritoneal dialysis for 12 years. Her history included frequent peritonitis.

In the early period, abdominal cocoon syndrome typically presents with nonspecific symptoms. Anorexia, nausea and intermittent abdominal pain may be present. In the early period, the physical examination is usually unremarkable. However, bloody dialysate effluent may be detected, especially during fluid exchange after periods when the peritoneal dialysis cavity is dry (8). In the later period, patients present with symptoms of ileus and peritoneal adhesion. Complaints of severe abdominal pain, vomiting and constipation are present due to small bowel obstruction (9). Accordingly, our case had complaints of nausea, vomiting and abdominal pain that had persisted for 3 months, was introduced

Table 1. Patient's Laboratory Parameters At Admission, On The 10th Day of Treatment and At 3 Months Post-Discharge

Parameter	At Initial Admission	10 th Day of Treatment	3 Months Post Discharge
WBC	12200 /ul	9880 /ul	8750 /ul
HGB	8.9 gr/dl	8.2 gr/dl	10.4 gr/dl
NA	136 mmol/l	138 mmol/l	136 mmol/l
K	3 mmol/l	5 mmol/l	5.5 mmol/l
CALCIUM	8.1 mg/dl	8 mg/dl	9 mg/dl
P	2.5 mg/dl	3.4 mg/dl	5.5 mg/dl
BUN	18 mg/dl	43 mg/dl	48 mg/dl
CREATININE	2.7 mg/dl	4.4 mg/dl	5.5 mg/dl
CRP	166 mg/L	37 mg/L	3 mg/L

to surgery due to presenting with a picture of ileus but the operation was terminated without performing a specific procedure due to the adhesions.

Laboratory results show non-specific findings such as a high white blood cell count. Radiologically, findings of peritoneal thickening, dilation of bowel loops and peritoneal calcification are significant. Characteristically, in the absence of symptoms, peritoneal calcification and bowel dilation serve as the most important findings (10). The CT of our patient showed dilation of bowel loops and fluid collections.

Medical or surgical treatment can be administered based on the clinical findings of the patient. At the initial stage, the patient should be commenced on bowel rest and nutritional support (11). Peritoneal resting was shown to alleviate the symptoms. The duration of this should vary between 4-12 weeks until the symptoms ameliorate. For this purpose, placing a central venous catheter and proceeding with hemodialysis would be an appropriate choice (12). Also, the patients should be carefully monitored with regard to malnutrition and switched to parenteral feeding when necessary. Our case also showed insufficient oral feeding. Laboratory parameters indicating nutrition were disturbed. Laboratory results at admission and post-discharge are presented in Table 1.

Recently, corticosteriods, tamoxifen and immunosupressive agents are being used in the treatment. These medications are reported to reduce peritoneal inflammation and fibrosis (13). Tamoxifen should be administered for 3 months at a daily dose of 10 mg. In addition, methylprednisolone should be given for 4 months at a dose of 40 mg, and then, should be ceased by tapering the dose over a period of 6-8 weeks (12). In a study that included 111 patients, patients receiving tamoxifen and patients not receiving tamoxifen were not significantly different with respect to mortality (13). The administered agent, duration of administration and the dose vary across different studies.

Surgical treatment is generally not recommended due to the possibility that the adhesions will worsen, as well as the acute obstruction occurring due to abdominal cocoon syndrome and perforation. Predominantly, medical treatment is recommended in chronic cases. However, if surgery is to be performed on chronic cases, it should be conducted at experienced centers (14). In our case, medical treatment could have been attempted prior to surgery had this disease been considered as a preliminary diagnosis. Clinical improvement began on only the 7th day of methylprednisolone therapy and clinical, laboratory findings completely recovered at the 3rd month of follow-up.

Abdominal cocoon syndrome is a rare disease, however, it reduces the quality of life and has a fatal progression. Surgery is often not recommended except for acute complications. Therefore, the medical treatment of the disease holds significance. In patients receiving long-term peritoneal dialysis who present with a picture of ileus, particularly if there have been frequent peritonitis episodes, abdominal cocoon syndrome should be considered in the differential diagnosis and timely medical treatment should be provided. If not treated timely and correctly, the disease can lead to complications such as sepsis, malnutrition and mortality. Here, we aimed to contribute to the literature with the treatment used in the presented case.

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