Teaching Point
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Fulminant sclerosing peritonitis immediately following acute bacterial peritonitis

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Abstract
Sclerosing peritonitis (SP) is an uncommon disease in the peritoneal dialysis population, but the considerable morbidity and mortality associated with it make early diagnosis and treatment imperative. We describe a case illustrating that a fulminant variant of SP can occur as a second phase phenomenon immediately following treatment of acute bacterial peritonitis, a setting in which the diagnosis may be masked and aggressive immunosuppression considered potentially hazardous. Treatment with corticosteroid alone appears particularly effective for SP occurring in this setting.

Keywords: CAPD peritonitis; immunosuppression; inflammation; sclerosing encapsulating peritonitis

Introduction
Sclerosing peritonitis (SP) is an uncommon disease in the peritoneal dialysis (PD) population [1], and is characterized by fibrosis of both the visceral and parietal peritoneum. The clinical presentation of SP is highly variable and, if unrecognized and untreated, it is associated with considerable morbidity and mortality.

We describe a 54-year-old female PD patient who, immediately following appropriate treatment of bacterial peritonitis, had an unrelenting acute inflammatory state with life-threatening anorexia and malnutrition. Despite numerous antimicrobial agents, her condition deteriorated and her survival was uncertain.

A fulminant variant of SP can occur as a second phase phenomenon immediately following treatment of acute bacterial peritonitis, but in this setting aggressive immunosuppression is potentially hazardous. A high degree of clinical acumen is necessary to recognize this form of the disorder, and initiate potentially life-saving treatment with corticosteroid monotherapy, which appears particularly effective for SP occurring in this setting.

Case
A 54-year-old female developed end-stage renal disease due to congenital renal dysplasia, and had been maintained on continuous ambulatory peritoneal dialysis (CAPD) for 10 years. During this period, there had been only two isolated episodes of peritonitis that settled rapidly with appropriate antibiotics. Right breast carcinoma, 5 years after commencement of dialysis, was managed by local excision and adjuvant radiotherapy. She had no other ill health.

She was hospitalized with a 24 h history of vomiting and abdominal pain. On examination, she had a fever of 38.7°C, dehydration, abdominal tenderness and rebound. Coliforms were grown on culture of peritoneal fluid. Failure to improve after 48 h of appropriate antibiotic therapy led to laparotomy and PD catheter removal. Haemodialysis therapy was commenced.

Despite these measures, right upper quadrant pain and fever persisted, together with hypoactive bowel sounds and a grossly elevated C-reactive protein (CRP), consistently >200 mg/L. Extensive microbiological cultures were negative and there was no response to numerous antimicrobial agents. There was an unrelenting fever, complete anorexia and rapid weight loss of 11 kg in 6 weeks.

Computed tomography (CT) scanning of the abdomen revealed a perihepatic fluid collection, and multiple small low-density lesions in the liver. Ascitic fluid aspirate was sterile on culture and did not show malignant cells.

In view of this clinical course, the operative findings at initial laparotomy were reviewed; the peritoneum had been described as thickened, sclerosed and densely adherent to viscera in the right upper quadrant.

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Oral prednisolone at a dose of 1 mg/kg daily was commenced. Within 12 h her temperature became normal, CRP fell from 180 mg/L to 29 mg/L by 72 h (Figure 1), and her anorexia resolved. Prednisolone was reduced to 0.3 mg/kg daily. After 12 days she was discharged with complete resolution of symptoms, no fever and a CRP of 11 mg/L.

Prednisolone therapy was slowly tapered and discontinued after 10 months. Eighteen months later, she remains well on regular haemodialysis therapy with no recurrence of symptoms.

Discussion

SP is characterized by fibrosis of the visceral and parietal peritoneum. It has been reported to occur in association with PD, certain β-blockers, ovarian tumours, connective tissue diseases and liver disease. (Primary SP principally remains a disease of adolescent females in the tropics and sub tropics.)

Injury to, or irritation of, the peritoneal mesothelium is considered to be the initial stimulus for SP in susceptible individuals. This is followed by florid reactive hyperplasia of the mesenchymal cells, producing a thick layer of fibroconnective tissue. In PD, the continual exposure to non-physiological fluid and intermittent bacterial infection are probable precipitants [2].

The presentation of SP is variable. The most common manifestations are ultrafiltration failure (in PD patients), anorexia, nausea and weight loss, or small bowel obstruction. These clinical findings reflect inhibition of peristalsis, fibrous tethering of bowel loops and encapsulating sclerosis (the ‘abdominal cocoon’). Ascites, which may be chylous in nature, develops when lymphatic drainage is impeded.

Initial radiological findings may be non-specific. CT scanning in advanced cases may reveal tethering of bowel loops and calcification [3]. These findings are variable, and diagnosis is most reliably made by laparotomy or laparoscopy.

SP has a poor outcome and therapy is not evidence based, reflecting the rarity of the diagnosis in the peritoneal dialysis population, estimated at 0.7% prevalence in one study [1].

Discontinuation of peritoneal dialysis and transfer to haemodialysis is considered mandatory. Supportive measures include nutritional support with enteral or parenteral supplementation. The role of surgical intervention is unclear [4].

On the basis of anecdotal reports of benefit with immunosuppressive agents, and the adverse prognosis without intervention, immunosuppressive therapy is now considered as first line treatment. Various regimes have been used. Anti-fibrotic agents, while conceptually attractive, have proved disappointing; tamoxifen and, more recently, sirolimus have been used without significant benefit [5,6]. In one case, there was no improvement with sirolimus monotherapy, but a rapid clinical improvement when a switch to corticosteroid monotherapy was initiated [6].

The case we report was characterized by a dramatic response to corticosteroid therapy, and also a delay in diagnosis due to the mode of presentation. On review of the literature, there have been four previously published case reports where the presentation of SP has been dominated by a persistent acute inflammatory state [5–8]. In none of these cases was the diagnosis immediately obvious, and there was a delay in commencement of appropriate therapy. In at least two cases, there was a striking response within a week to corticosteroid monotherapy [5,8].

In the case series reported in the literature, the presenting features of each case are not always documented. In those that are, intestinal obstruction is undoubtedly the most common presenting feature. Severe preceding infective peritonitis is reported [1,9,10], but the temporal proximity of the peritonitis episode to the diagnosis of SP is uncertain.

In the Australian study [1], it seems likely that there were cases with similar clinical features to our case report, (‘[in the patients with peritonitis] the bowel function often did not recover and the patient may have died from on-going sepsis’), but crucially without recognition that the persistent inflammatory state may have been due to SP rather than on-going sepsis.

In the only published prospective study of SP [9], there were ‘12 patients (25%) in whom bacterial peritonitis was thought to directly trigger the onset of EPS’. There is no additional information on this group of patients or their management. Of interest, in the Korean study of 34 cases published earlier this year, steroid therapy was not utilized in the management of any patient [10]. The therapy was total parenteral nutrition ± surgery.

The literature is insufficiently developed to allow a discernible difference in response to immunosuppressive therapy between the indolent form of SP (presenting with ultrafiltration failure and abdominal pain) and the fulminant form (characterized by a
persistent acute inflammatory state after peritonitis) to be determined. The clinical information in the case series is limited, and the case reports with immunosuppression vary in drug chosen, dose and duration of therapy, clinical features of the cases and the initiation of pharmacological therapy in the time course of the disease process. Future studies should seek to address this issue, given the relevance to both the pathogenesis and treatment options of establishing if there is a clinical distinction between the indolent form of SP and that presenting after infective peritonitis.

This case highlights the importance of considering the diagnosis of SP in PD patients with apparent non-resolution of bacterial peritonitis despite appropriate treatment. It is an uncommon but potentially fatal condition if untreated, and the diagnosis requires a high index of clinical suspicion.

Corticosteroid treatment appears particularly effective when the condition is characterized by an acute inflammatory state, and is similar in this respect to the retroperitoneal fibrosis that affects some patients with an inflammatory abdominal aortic aneurysm.

**Teaching points**

In conclusion, we feel that this case and review of the contemporary literature convey important learning points for nephrologists.

1. Acute bacterial peritonitis may occasionally trigger a persisting intra-abdominal inflammatory syndrome of autoimmune nature.
2. This manner of presentation of SP may be misinterpreted as unresolved sepsis and may lead to a potentially fatal delay in diagnosis.
3. A literature review of this area suggests that it is not well recognized that SP may present in this way, and represent a fulminant variant of the disorder.

4. Recognition of the correct diagnosis and corticosteroid treatment may be life saving in these circumstances.

**Conflict of interest statement.** None declared.

**References**


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