Case

In June 2000 a 36-year-old, male chronic haemodialysis patient was transferred to our hospital with gradual weight loss of 7 kg during 7 months (body mass index decrease from 19.5 to 17), abdominal pain and loss of appetite with resulting cachexia. On palpation, tenderness and guarding in all abdominal quadrants were noted, on auscultation bowel sounds were normal without tinkling. A plain abdominal X-ray (Figure 1) revealed gas and fluid levels in the ileum suggesting paralytic or obstructive ileus. In addition the patient complained of massive arthralgias of both hips, knees, and ankles which developed 2 days prior to admission, together with maculopapular efflorescences on both lower limbs. Laboratory evaluation at admission is shown in Table 1.

In 1991 ESRD had been diagnosed without establishing the underlying renal disease. After being treated with CAPD for 4 months the patient received a renal allograft, which had functioned for about 3 years before graft function decreased due to chronic graft nephropathy. CAPD treatment was resumed for another 4.5 years. He suffered from multiple episodes of tunnel infections with resulting peritonitis. In November 1999 CAPD had to be abandoned due to progressive ultrafiltration failure and the patient was started on intermittent haemodialysis treatment. In February 2000 the CAPD catheter was removed. Since November 1999 the patient had suffered from abdominal discomfort and distension, nausea, and vomiting. Ascites was shown by ultrasound to persist even 6 months after discontinuation of peritoneal dialysis.

Questions

What is the diagnosis?
What further investigations should be performed?
What treatment should be recommended?

Table 1. Laboratory results on admission

<table>
<thead>
<tr>
<th>Test</th>
<th>Value (mg/dl)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Creatinine</td>
<td>7.8</td>
</tr>
<tr>
<td>BUN</td>
<td>30.4</td>
</tr>
<tr>
<td>Albumin</td>
<td>3.2</td>
</tr>
<tr>
<td>CRP</td>
<td>9.3</td>
</tr>
<tr>
<td>Cholesterol</td>
<td>106</td>
</tr>
<tr>
<td>LDL chol.</td>
<td>44</td>
</tr>
<tr>
<td>Leukocytes</td>
<td>7,500</td>
</tr>
<tr>
<td>Immunglobulin-A</td>
<td>1,140</td>
</tr>
</tbody>
</table>

Fig. 1. Abdominal X-ray, taken in left lateral position on admission. Distended small intestine; fluid–gas levels especially in the terminal ileum and colon ascendens.
Answer to the quiz on preceding page

We diagnosed our patient as having sclerosing encapsulating peritonitis (SEP). This was suspected because of the history of multiple episodes of peritonitis and consecutive ultrafiltration loss, ascites, and the described obstructive symptoms. The papules macroscopically resembled Henoch–Schönlein purpura (HSP) lesions, which could also have been responsible for the abdominal and arthritic symptoms. We therefore arranged a skin biopsy, which revealed leukocytoclastic vasculitis of subcutaneous vessels, thus confirming the diagnosis. A computed tomography scan of the abdomen (Figure 2) showed the known intraabdominal fluid, intestinal distension, and coprostasis and wall thickening of the small intestines. Later we learned of a peritoneal biopsy taken at the time of peritoneal dialysis (PD) catheter removal showing severe sclerosis underlying the mesothelium.

Systemic steroids (1 mg/kg/day prednisolone equivalent with tapering after 6 weeks) were started as treatment for HSP and sclerosing peritonitis and led to a rapid improvement of the purpura. Because of the young age of the patient, we experimentally established tamoxifen as a potential antifibrotic agent [1]. During his hospital stay the patient was unable to eat because of intractable nausea, intra-abdominal discomfort, and vomiting shortly after ingestion. Therefore we decided to implant a central venous port system for total parenteral nutrition. Six months after initiation of this therapy the general condition of the patient improved and he gained 2 kg of body weight. Abdominal symptoms disappeared gradually and he started to eat small portions while parenteral nutrition was being reduced.

Fig. 2. Computed tomography of the abdomen with oral and intravenous contrast, taken on admission: Gross amounts of intraabdominal fluid, thickened parietal and visceral peritoneum, and evidence of atonic bowel.

Discussion

SEP is a rare event, affecting 0.5 to 0.9% of all PD patients [2]. It seems to be the maximal variant within a wide range of histological alterations of the peritoneum, beginning with simple sclerotic lesions. Histologically it is characterized by changes of the mesothelial cell layer, with loss of cell attachment to the basal membrane, duplication of the basal membrane, submesothelial oedema and thickening, combined with inflammatory infiltration, calcification, and vascular occlusion with intimal thickening and media hyalinization. Typically the thickness of the peritoneum in SEP exceeds 40 μm [3]. Macroscopically the sclerotic peritoneum could eventually lead to fixation and obstruction of intraperitoneal organs.

Significant sclerosis of the peritoneum is associated with bioincompatibility of PD solutions due to hyperosmolarity, high glucose content, acidity, acetate buffer, disinfectants, and foreign bodies, for example small plastic particles shed by tubes or fluid containers [3]. However, the most commonly described pathogenetic factor is peritonitis with mesothelial cell loss, intraperitoneal fibrin production, and loss of mesothelial fibrinolytic capacity [4,5].

Clinically, patients with SEP can, but need not necessarily, present with loss of ultrafiltration capacity together with abdominal symptoms, as has been seen in our patient. Also abdominal masses and a haemorrhagic dialysate can be present. Onset of symptoms can be slow or very acute. Diagnostic procedures consist of ultrasound examination, abdominal X-ray with or without contrast media, and computed tomography. A decrease of CA 125 in the effluent may be used as marker for diminishing mesothelial cell mass [6].

Therapeutic principles used are the discontinuation of PD treatment and removal of the catheter, treatment with immunosuppressive and antifibrotic agents such as steroids, azathioprine, colchicine, and even cyclophosphamide and experimentally also with gestagens and tamoxifen. Surgery should be reserved for acute intestinal obstruction, but removal of membranes is associated with a high rate of morbidity and mortality. Total parenteral nutrition is a valuable alternative for progressive malnutrition [5].

References

Progressive anorexia and chronic ascites after termination of CAPD
