Idiopathic Sclerosing Peritonitis in a Man

Chiyohiko Masuda, Yoshiaki Fujii, Tomoyoshi Kamiya, Matakihi Miyamoto, Katsuhiro Nakahara, Shigeo Hattori, Hisataka Ohshita, Takeshi Yokoyama, Hiroyuki Yoshida* and Yutaka Tsutsumi**

Idiopathic sclerosing peritonitis (ISP) is a rare condition which has been mainly reported in young adolescent women as a cause of small bowel obstruction. In these patients the small bowel is sometimes encased in a fibrous sac called an “abdominal cocoon”. We describe a 62-year-old man who underwent exploratory laparotomy for ascites and abdominal mass. Laparotomy showed 5.4 l of ascites and the entire small bowel was encased in a fibrous sausage-like cocoon. The pathological findings were characteristic of sclerosing peritonitis.

Key words: sclerosing peritonitis, abdominal cocoon

Introduction

Sclerosing peritonitis is a rare disease characterized by fibrosis and adhesion of the peritoneum to loops of the small intestine (1). It has been most commonly reported after treatment with β-adrenergic blocking agents (2–5), continuous ambulatory peritoneal dialysis (6), peritoneo-venous or ventriculo-peritoneal shunting (7–9). Idiopathic sclerosing peritonitis (ISP) has been described mainly in young adolescent women, especially those residing in subtropical climates. The small bowel is sometimes encased in a fibrous sac described as “abdominal cocoon”. The clinical picture usually includes repeated episodes of small bowel obstruction and abdominal mass. Recently a few cases with ISP in middle aged women or men have been reported (10, 11). Here we report a 62-year-old Japanese man with ISP.

Case Report

A 62-year-old man was admitted to our hospital because of abdominal fullness. The patient was well until about 1 year earlier, when he began to feel abdominal distention. He had no history of small bowel obstruction and has never taken β-adrenergic blocking agents. Physical examination showed a marked abdominal distention with a sign of ascites and a nontender rounded soft mass in the mid-abdomen. Computed tomography of the abdomen and barium studies revealed massive ascites and oval shaped mass in which small bowel was matted together (Fig. 1). Abdominal paracentesis fluid was clear containing 100 WBC/mm³ (70% lymphocyte), 4.2 g protein/100 ml and 250 IU LDH/l. Cytologic studies were negative for malignant cells. Cultures of ascitic fluid were negative for bacteria, fungi and tubercule bacilli. There was no evidence of hepatic, renal or cardiac disease. He underwent exploratory laparotomy. The laparotomy showed 5.4 l of ascitic fluid and fibrin in the pelvis. The entire small bowel was encased in a large sausage-like sac composed of a thick fibrous peel (Fig. 2). The small bowel could be seen entering this sac 20 cm distal to the ligament of Treitz and exiting just proximal to the ileocecal valve. The sac was attached to the root of the mesentery of the small bowel with no adhesion to the parietal peritoneum. The parietal layer of the peritoneum was thickened with white fibrous plaque. The spleen was covered by white fibrous membrane. Multiple biopsies were done. Pathological examination of biopsy specimens showed thick scaring fibrosis with scattered foci of perivascular inflammatory infiltration. The peritoneal surfaces were covered with a thick fibrous layer (Fig. 3). There was no evidence of tuberculosis, Crohn’s disease, or other granulomatous conditions. Neither neoplastic lesion nor artificial materials were seen in the specimens.

Discussion

Sclerosing peritonitis is a rare disease characterized by fibrosis and adhesion of the peritoneum, sometimes forming a sac in which the small bowel is totally or partially encased. Most
Idiopathic Sclerosing Peritonitis

Figure 1. Computerized tomography of the abdomen (left). Barium examination of the small bowel (right). Barium-filled ileal loops are matted together in mid abdomen with ascites.

Figure 2. The entire abdominal cocoon at laparotomy. The small bowel was entering the sausage-like cocoon 20 cm distal of the ligament of Treitz and exiting just proximal to the ileocecal valve. The peritoneal surfaces are covered with a thick fibrous layer.

Figure 3. Peritoneal biopsy specimens show thick scarring fibrosis with foci of perivascular inflammatory infiltration (HE stain, x20).

Cases were reported secondary to administration of β-adrenergic blocking agent, chronic peritoneal dialysis and peritoneovenous or ventriculo-peritoneal shunting. ISP was first described by Foo et al (1) in ten young adolescent girls in Singapore; in these cases the small bowel was encased by a thick fibrous membrane described as an "abdominal cocoon".

Twenty-one cases including the present case with ISP have been reported previously and summarized in Table 1. The mean age of the patients was 27 years (range 4–62 years). Seventeen were female and four were male. Fifteen of the seventeen female patients were young adolescent girls. All of the male patients have been reported more recently and tend to be older than the female patients. The small bowel obstruction was noted in 16 cases and ascites has been reported in 4 previous cases. The ascites in the present case was exudate. All cases except for ours resided in subtropical climates. The present case is the first description of this condition in an oriental man.

Severe adhesion and fibrosis of the peritoneum with ascites can occur after abdominal surgery and pyogenic and tubercu-
Table 1. Cases of Idiopathic Sclerosing Peritonitis

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>Number of patients</th>
<th>Age</th>
<th>Sex</th>
<th>Number with small bowel obstruction</th>
<th>Number with ascites</th>
<th>Geographical distribution</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Foo et al</td>
<td>1978</td>
<td>10</td>
<td>13-18</td>
<td>F</td>
<td>8</td>
<td>0</td>
<td>Singapore</td>
<td>1</td>
</tr>
<tr>
<td>Rao et al</td>
<td>1979</td>
<td>1</td>
<td>4</td>
<td>F</td>
<td>1</td>
<td>0</td>
<td>India</td>
<td>12</td>
</tr>
<tr>
<td>Sayfan et al</td>
<td>1979</td>
<td>1</td>
<td>12</td>
<td>F</td>
<td>1</td>
<td>0</td>
<td>Israel</td>
<td>14</td>
</tr>
<tr>
<td>Marinho et al</td>
<td>1980</td>
<td>1</td>
<td>17</td>
<td>F</td>
<td>0</td>
<td>0</td>
<td>Nigeria</td>
<td>16</td>
</tr>
<tr>
<td>Sieck et al</td>
<td>1983</td>
<td>1</td>
<td>14</td>
<td>F</td>
<td>1</td>
<td>1</td>
<td>Saudi Arabia</td>
<td>13</td>
</tr>
<tr>
<td>Dehn et al</td>
<td>1985</td>
<td>1</td>
<td>12</td>
<td>F</td>
<td>1</td>
<td>0</td>
<td>UK</td>
<td>15</td>
</tr>
<tr>
<td>Narayanan et al</td>
<td>1989</td>
<td>4</td>
<td>12,40</td>
<td>F</td>
<td>1</td>
<td>1(200 ml)</td>
<td>India</td>
<td>10</td>
</tr>
<tr>
<td>Narayanan et al</td>
<td>1989</td>
<td>4</td>
<td>40,60</td>
<td>M</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Burstein et al</td>
<td>1990</td>
<td>1</td>
<td>35</td>
<td>M</td>
<td>1</td>
<td>0</td>
<td>Israel</td>
<td>11</td>
</tr>
<tr>
<td>Present case</td>
<td>1992</td>
<td>1</td>
<td>62</td>
<td>M</td>
<td>0</td>
<td>1(5.4 l)</td>
<td>Japan</td>
<td>Present case</td>
</tr>
</tbody>
</table>

Although there was no evidence of retroperitoneal fibrosis in this case, these facts are of interest in that ISP may share a common etiology with idiopathic retroperitoneal fibrosis. To elucidate the precise etiology of ISP, further study of cases is necessary.

References

12) Rao PLNG, Mitra SK, Pathak IC. Abdominal cocoon – a cause of...