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SOLITARY FIBROUS TUMOR IN THE RETROPERITONEUM: A CASE WITH INFILTRATIVE GROWTH

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Solitary fibrous retroperitoneal tumor is rare. We present a case with infiltrative growth in a 56-year-old female patient whose initial symptom was palpable tumor in the lower abdomen. Computed tomography and magnetic resonance imaging indicated a mass in the retroperitoneum under the left kidney with a poorly demarcated infiltrative growth. Surgical findings revealed a gelatinous tumor in the retroperitoneum, which had invaded up to the fatty tissue surrounding the Gerota's fascia and to the fatty tissue surrounding the descending colon. However, as there was no invasion into the Gerota's fascia, it was possible to preserve the left kidney. Pathohistological examination revealed increased cellularity in the tumor tissues as well as tissues with atypical nuclei of the tumor cells with some cell division. Due to these findings, it was diagnosed as malignant solitary fibrous tumor. Only surgical treatment was performed and the patient is alive without recurrence 2 years and 4 months after surgery.

Key words: Solitary fibrous tumor, Retroperitoneum, Malignant tumor, Infiltrative growth

INTRODUCTION

Solitary fibrous tumor (SFT) was first reported in 1931 as a primary neoplasm of the pleura. It is a rare spindle cell tumor that occurs in adults and is known as a benign tumor of the pleura often described as encapsulated or sharply circumscribed. It rarely occurs in the retroperitoneum, and only 24 cases have been reported in the English-language literature. Among them, only 2 cases were malignant SFT, and our case is the first case of retroperitoneal SFT with infiltrative growth in the surrounding tissues.

CASE REPORT

The patient was a 56-year-old female who noticed a mass in the left lower abdomen and visited our hospital. On physical examination, a hard mobile mass was palpable in the left lower abdomen. Values of carcinoembryonic antigen, carbohydrate antigen 19-9 and cancer antigen 125 were normal. Excretory urography indicated no abnormality in the urinary tract. Barium enema revealed a compression figure in the descending colon. Abdominal ultrasonography demonstrated a somewhat poorly demarcated tumor with heterogenous internal echo. Computed tomography (CT) revealed a slightly heterogenous 9×6×6 cm tumor in the retroperitoneum under the left kidney. Enhanced CT showed a tumor with some deep staining. Magnetic resonance imaging (MRI) T1-weighted image were low intensity similar to the muscles (Fig. 1a), but T2-weighted images were high intensity and internally partly heterogenous (Fig. 1b). The tumor was poorly demarcated with a bean-like component extending posteriorly, closely bordering the intestines and expanding up along the Gerota's fascia. CT as well as MRI revealed no lymph node metastasis around the aorta or hepatic metastasis. Angiography showed an internally hypovascular tumor with an indistinct outline, but no feeding vessels. Malignant teratoma or sarcoma occurring in the retroperitoneum was suspected and laparotomy was performed. On laparotomy, a partly encapsulated tumor was observed and in the retroperitoneum, a poorly demarcated gelatinous substance had spread diffusely up to the fatty tissue surrounding the Gerota's fascia and the fatty tissue surrounding the descending colon. The tumor was removed along with the fatty tissue in the retroperitoneum containing the gelatinous substance, and part of the descending colon, and the lower half of the Gerota's fascia were excised as well. In the removed specimen, the encapsulated part of the tumor was filled with a gelatinous substance, but there were no necrotic tissues. Pathohistological examination revealed weakly amplified images with varied cellular intensity, remarkable collagen fiber hyperplasia in the interstitium and some hyalinization. Invasion into the surrounding fatty tissues was also observed (Fig. 2a), and abundant bean-like hemangiopericytoma blood vessels were
Abdominal MRI. (a) T1-weighted transverse image reveals a tumor (black arrow heads) with a beak-like component extending posteriorly into the fatty tissue and closely bordering the intestines; (b) T2-weighted coronal oblique image shows the high intensity tumor extending up along the Gerota's fascia.

Fig. 1. Abdominal MRI. (a) T1-weighted transverse image reveals a tumor (black arrow heads) with a beak-like component extending posteriorly into the fatty tissue and closely bordering the intestines; (b) T2-weighted coronal oblique image shows the high intensity tumor extending up along the Gerota's fascia.

observed (Fig. 2b). There were both tissues with weak and strong nuclear atypia, and strongly magnified images of the tissue with strong nuclear atypia revealed cells with spindle shaped nuclei and nuclear atypia with some cell division (1 mitoses/10 HPFs) (Fig. 2c). Immunohistological staining indicated the presence of vimentin and CD34 and absence of S-100, actin and CD68. Accordingly, a diagnosis of retroperitoneal malignant SFT was made. No further treatment was performed, and the patient is alive without recurrence 2 years 4 months after surgery.

DISCUSSION

SFT originating in the pleura has been well documented\(^{14}\), and cases of extrapleural SFT have also been reported. When the 25 reported cases of retroperitoneal SFT including our present case (Table 1) were examined, there were 11 male and 13 female patients aged 17 to 82 (mean 49.0), with tumor sizes from 2 to 26 cm (mean 10.6 cm). The symptoms in 8 cases were incidental, followed by pain and hypoglycemia in 3 cases. Histologically, 21 were benign and 3 were malignant. Excluding our case, all others were well circumscribed or encapsulated, and ours was the first case of retroperitoneal SFT with infiltrative growth. Surgical excision was performed
Table 1. Cases of retroperitoneal solitary fibrous tumor

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Size (cm)</th>
<th>Symptom</th>
<th>Histology</th>
<th>Margin</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nevius</td>
<td>1959</td>
<td>48</td>
<td>M</td>
<td>ND</td>
<td>hypoglycemia</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>Echenique</td>
<td>1988</td>
<td>58</td>
<td>M</td>
<td>14</td>
<td>voiding difficulty</td>
<td>benign</td>
<td>completely encapsulated</td>
<td>SE+radiation</td>
<td>NED</td>
</tr>
<tr>
<td>Young</td>
<td>1990</td>
<td>59</td>
<td>F</td>
<td>9</td>
<td>incidental</td>
<td>benign</td>
<td>well-circumscribed</td>
<td>SE</td>
<td>NED</td>
</tr>
<tr>
<td>Goodlad</td>
<td>1991</td>
<td>66</td>
<td>M</td>
<td>7</td>
<td>frequency, nocturia</td>
<td>benign</td>
<td>well-circumscribed</td>
<td>SE</td>
<td>NED</td>
</tr>
<tr>
<td>Piazza</td>
<td>1996</td>
<td>51</td>
<td>F</td>
<td>7</td>
<td>incidental</td>
<td>benign</td>
<td>well-circumscribed</td>
<td>SE</td>
<td>NED</td>
</tr>
<tr>
<td>Fukunaga</td>
<td>1997</td>
<td>33</td>
<td>F</td>
<td>3.5</td>
<td>ND</td>
<td>benign</td>
<td>well-circumscribed</td>
<td>SE</td>
<td>NED</td>
</tr>
<tr>
<td>Nielsen</td>
<td>1997</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>malignant</td>
<td>well-circumscribed</td>
<td>SE</td>
<td>ND</td>
<td>NED</td>
</tr>
<tr>
<td>Decouvelaere</td>
<td>1998</td>
<td>40</td>
<td>M</td>
<td>17</td>
<td>ND</td>
<td>benign</td>
<td>well-circumscribed</td>
<td>SE</td>
<td>lung metastasis</td>
</tr>
<tr>
<td>Yokoi</td>
<td>1998</td>
<td>38</td>
<td>M</td>
<td>ND</td>
<td>hypoglycemia</td>
<td>malignant</td>
<td>well-circumscribed</td>
<td>SE</td>
<td>local recurrence</td>
</tr>
<tr>
<td>Hasegawa</td>
<td>1999</td>
<td>48</td>
<td>F</td>
<td>5</td>
<td>incidental</td>
<td>benign</td>
<td>well-circumscribed</td>
<td>SE</td>
<td>NED</td>
</tr>
<tr>
<td>Morimatsu</td>
<td>2000</td>
<td>82</td>
<td>M</td>
<td>23</td>
<td>ND</td>
<td>benign</td>
<td>well-circumscribed</td>
<td>SE</td>
<td>NED</td>
</tr>
<tr>
<td>Clayton</td>
<td>2001</td>
<td>65</td>
<td>M</td>
<td>5</td>
<td>incidental</td>
<td>benign</td>
<td>ND</td>
<td>biopsy</td>
<td>no change</td>
</tr>
<tr>
<td>Present case</td>
<td>56</td>
<td>F</td>
<td>9</td>
<td>abdominal mass</td>
<td>malignant infiltrative</td>
<td>SE</td>
<td>NED</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ND: not described, SE: surgical excision, NED: alive with no evidence of disease.

in 21 cases, and surgical excision and radiation in 2 cases. As for the prognosis, 20 cases had no evidence of disease, 2 cases were alive with disease and 1 case had lung metastasis. These results were compared with those of 223 cases of pleural-based SFT reported by England et al.\(^4\) Although the age was 8 years younger for retroperitoneal SFT, sex, tumor size and symptoms were similar. The ratio of malignant SFT was 37% for pleural-based SFT, while it was 12.5% for retroperitoneal SFT. As for prognosis, recurrence was observed in 23% and metastasis in 9.8% for pleural-based SFT, while both recurrence and metastasis were 4.2% for retroperitoneal SFT. These results suggest that retroperitoneal SFT may be histologically and clinically more benign compared to pleural-based SFT.

Histological malignancy is unlike biological malignancy, and there have been reports of metastasis even in benign SFT\(^6\). Even if it were malignant, complete excision of the tumor is thought to have a favorable prognosis, and resectability is the most important indicator of clinical outcome. In our case, a poorly demarcated gelatinous substance had spread invasively into the surrounding tissues, making it difficult to decide the extent of excision.

Solitary fibrous tumor of the retroperitoneum is rare, and in particular, no reports have been published on retroperitoneal solitary fibrous tumors with infiltrative growth. In our case, although the tumor was histologically malignant and infiltrative, the outcome of surgical treatment without further treatment was good. The patient is without recurrence at 2 years 4 months after surgery. However, due to reported cases of recurrence after surgical excision, this case is being carefully followed on a long-term basis.

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浸潤性発育を示した後腹膜孤立性線維性腫瘍の1例

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孤立性線維性腫瘍は稀な疾患である。今回われわれは56歳の女性に発生した悪性後腹膜孤立性線維性腫瘍の1例を報告した。主訴は腹部腫瘤触知で、腹部 CT および MRI 検査にて左腎下方向に接して、Gerota 筋膜に沿うように発育する境界不明瞭な腫瘍を認めた。手術所見では後腹膜腔にゼラチン状の腫瘍を認め、下行結腸周囲の脂肪組織や Gerota 筋膜周囲の脂肪組織に浸潤していた。Gerota 筋膜内への浸潤は認めなかったので左腎は温存した。病理組織像では類質の線維腫瘍組織が著明に増殖し、hemangiopericytoma 様のくぼみ状を呈する血管も豊富に認められた。腫瘍組織内に腫瘍細胞密度の増加しているところや核異型度が強く分裂像を認める組織が混在しており、また周囲脂肪組織内に浸潤している所見を認めた。以上より、悪性後腹膜孤立性線維性腫瘍と診断した。治療は外科的切除のみを行い、2年4カ月を経過しても再発無く生存中である。

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