Carcinosarcoma arising from atypical endometriosis in a cesarean section scar

J. LENG*, J. LANG*, L. GUO†, H. LI* & Z. LIU*
Departments of *Gynecology and Obstetrics and †Pathology, Peking Union Medical College Hospital, CAM&PUMC, Beijing, China


Malignant change of endometriosis in a cesarean scar (CS) is rare. We report a case of carcinosarcoma arising from atypical endometriosis in a CS scar, which was successfully treated with complete excision of the lesion and repair of the abdominal wall defect with autologous skin–muscle flap graft. A 41-year-old woman presented with a recurrent endometriosis in a CS scar. Within 16 years it changed from benign to atypical endometriosis and finally to carcinosarcoma after three operations. Complete excision of the tumor was performed, with a big defect of abdominal wall successfully repaired by autologous pedicle skin–muscle graft. The diagnosis of carcinosarcoma arising from atypical endometriosis was confirmed histologically. The lesion recurred 6 months after the fourth operation. She died of disease 15 months after the fourth operation. This case demonstrated that long-standing recurrent scar endometriosis could undergo malignant changes and should be made aware. The primary treatment is complete surgical excision.

KEYWORDS: atypical endometriosis, carcinosarcoma, endometriosis, malignant change.

Endometriosis is a common disease that involves up to 10–15% of women in the reproductive age group. It is most frequently detected in the pelvic organs. Extrapelvic endometriosis is a fairly uncommon event. The majority of extrapelvic endometriosis involves scar tissues following obstetric and gynecologic procedures.
Cesarean section scar endometriosis is the most common form of extrapelvic endometriosis. Although endometriosis is a benign disease, it has been reported that some ovarian cancer arose from ovarian endometriosis and the malignant change occurred in about 1% cases of ovarian endometriosis\(^1\). Malignant change developing in extrapelvic endometriosis is rare, with the majority of cases being endometrioid adenocarcinoma and clear cell carcinoma\(^2,3\). We report a case of carcinosarcoma arising from atypical endometriosis in an abdominal scar and application of autologous skin–muscle flap to repair the abdominal wall defect after complete excision. To our knowledge this is the first such reported case.

Case report

A 41-year-old woman, grava 2, para 1, presented with a big painful abdominal mass, which was gradually enlarging after three previous excisions of an endometriotic nodule in a cesarean section scar. The pathology of nodule was reported as “atypical endometriosis.” This patient had a 16-year history of recurrent cesarean scar (CS) endometriosis. Sixteen years ago, she noticed a small mass of about 1 cm in abdominal scar 50 days after her CS. The size of the mass remained unchanged, and she only experienced mild pain at the wound during menstruation. She did not visit doctor until 12 years later when the mass had enlarged to 3 cm and the pain became cyclic and severe just prior to menstruation. The lesion was excised and was diagnosed as scar endometriosis histologically. Two years after the initial operation, she noticed a recurrent mass in the CS scar. It enlarged rapidly to 5 cm in diameter and was again removed surgically, with a diagnosis of recurrent scar endometriosis histologically. One month later, a similar mass of 7 cm recurred at the same scar site. She was given Gestrinone treatment for 3 months, but the mass continued to grow. She was then referred to our tertiary hospital for further management. At this time, physical examination revealed a partly solid and partly cystic tumor of 8 × 10 cm at right side of the abdominal scar. Pelvic examination showed a bulky uterus, normal-sized ovaries, and normal finding of whole abdominal and pelvic cavity exploration. A review of the histology of both the first and second excised specimens confirmed the diagnosis of scar endometriosis, but with features of atypical endometriosis. A biopsy from the tumor confirmed the same pathology of atypical endometriosis. Gross complete excision with surgical margin of 0.5 cm beyond the lesion of the abdominal wall including the right rectus abdominis muscle, together with a laparoscopic hysterectomy/bilateral salpingo-oophorectomy, was performed. At the time of operation, there was no pelvic endometriosis or ovarian endometriosis. Exploration of whole abdominal and pelvic cavity was normal. The pathology showed a small fibroid in the uterus, and no abnormality in both ovaries was found. Pathologic examination of resected abdominal wall lesion showed atypical endometriosis with clear resection margins, and pathologic minimal distance from the lesion of atypical endometriosis was 0.1 mm. The specimen showed negative estrogen and progesterone receptors. The patient was not followed up until 9 months later when a tumor recurred at the same abdominal wall scar. At examination, the lesion was irregular, bluish, solid-cystic, and tender, with ulceration of the skin in the right side of abdominal wall (Fig. 1). A biopsy from the tumor confirmed the diagnosis of malignancy. X-ray examination showed normal chest. Ultrasound scan showed normal liver, spleen, pancreas, kidneys, and no mass in the abdominal cavity. Wide radical resection of abdominal mass was performed with 1 cm beyond the visible lesion margin. The excised tumor consisted of fine cysts with dark-brown fluid and papillary friable necrotic tissues and had invasion into the rectus sheath, rectus muscle, external and internal oblique muscles, and peritoneum. Complete removal of the tumor created a large defect of 20 × 16 cm, which was repaired with a pedicle skin–muscle flap from the lateral side of the right thigh and a free skin flap from the left thigh. The patient had an uneventful recovery. The histologic examination revealed endometrioid carcinoma and sarcoma arising in areas of atypical glands and stroma of endometriotic lesion in the resected abdominal wall lesion (Fig. 2). The margins were not clear with infiltration of atypical endometriosis.

Figure 1. Abdominal lesion showed an irregular solid cystic mass with ulceration.
Immunohistological staining showed positive epithelial membrane antigen (EMA) and p53 expression and negative progesterone receptor and estrogen receptor expression in the epithelial component, whereas positive Vimentin and smooth muscle antigen (SMA) expression in stromal component. One course of chemotherapy with cisplatin and ifosfamide was given 7 days postoperatively and then discontinued because of hematologic toxicity. Two months after operation, a small mass of about 1 cm was found beyond the skin graft region and it was again excised, with a histologic diagnosis of atypical endometriosis. The patient was alive 12 months after the last operation, with recurrent small mass at the scar. She refused further operation and sought treatment with Chinese medicine. She died of the disease 15 months after the fourth operation due to pulmonary metastasis.

The patient had been living in rural area in the northeast of China since birth, and she had no history of other medical disorders.

**Discussion**

Surgical excision is the primary treatment for scar endometriosis, and complete removal of the lesion is important to prevent recurrence. Our patient presented with recurrent abdominal scar endometriosis, and after 16 years, it had a malignant transformation from benign to atypical endometriosis and finally to carcinosarcoma and endometrioid carcinoma after four surgical operations. Sampson first described malignant change in endometriosis in 1925. He proposed the criteria for diagnosis of carcinomatous development in endometriosis. These were 1) the coexistence of carcinoma and endometriosis in the same ovary; 2) a similar histologic relationship to each other; and 3) the exclusion of a second malignant tumor elsewhere. Later, in 1953, Scott postulated that the morphologic demonstration of benign endometriosis contiguous with the malignant tissue is prerequisite for adjudication of a malignancy originating in endometriosis in addition to the criteria by Sampson. The case we presented fulfilled these criteria. Malignant change in endometriosis most commonly develops within the ovary. However, 20% of malignancy in endometriosis occurs in extragonadal sites. The majority of malignant change of scar endometriosis is clear cell carcinoma or endometrioid carcinoma. A literature search in the Medline did not show any report on malignant change of stroma. Previous reports on malignant change from scar endometriosis are few and are shown in Table 1. The case we presented is the first report on carcinosarcoma arising from atypical endometriosis in a scar endometriosis.

The pathogenesis for the malignant change of endometriosis is not clear. But some studies suggest that certain aspects of endometriosis are similar to those of malignant disease. For example, like cancer, endometriosis can be both locally and distantly metastatic; it

**Table 1. Cases of malignant change arising in scar endometriosis**

<table>
<thead>
<tr>
<th>Author</th>
<th>Patient age (years)</th>
<th>Scar</th>
<th>Histology</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Madsen et al. (6)</td>
<td>49</td>
<td>Abdominal</td>
<td>Endometrioid carcinoma</td>
<td>Excision/castration/XRT</td>
<td>NA</td>
</tr>
<tr>
<td>Hitti et al. (7)</td>
<td>46</td>
<td>CS</td>
<td>Clear cell carcinoma</td>
<td>TAH/BSO/excision</td>
<td>NED at 30 months</td>
</tr>
<tr>
<td>Hitti et al. (7)</td>
<td>43</td>
<td>Episiotomy</td>
<td>Clear cell carcinoma</td>
<td>Chemo/XRT</td>
<td>DOD at 30 months</td>
</tr>
<tr>
<td>Miller et al. (3)</td>
<td>38</td>
<td>Abdominal</td>
<td>Clear cell carcinoma</td>
<td>Excision/chemo/XRT</td>
<td>NED at 60 months</td>
</tr>
<tr>
<td>Markopoulos et al. (9)</td>
<td>50</td>
<td>CS</td>
<td>Endometrioid carcinoma</td>
<td>Excision</td>
<td>NED at 24 months</td>
</tr>
<tr>
<td>Gucer et al. (2)</td>
<td>45</td>
<td>CS</td>
<td>Endometrioid carcinoma</td>
<td>Excision</td>
<td>NA</td>
</tr>
<tr>
<td>Park et al. (8)</td>
<td>54</td>
<td>CS</td>
<td>Clear cell carcinoma</td>
<td>Excision</td>
<td>NA</td>
</tr>
<tr>
<td>Matter et al. (10)</td>
<td>60</td>
<td>CS</td>
<td>Cystadenocarcinoma</td>
<td>Excision</td>
<td>NED at 12 months</td>
</tr>
<tr>
<td>Leng (this study)</td>
<td>41</td>
<td>CS</td>
<td>Carcinosarcoma</td>
<td>Excision</td>
<td>DOD at 15 months</td>
</tr>
</tbody>
</table>

XRT, radiotherapy; TAH, total abdominal hysterectomy; BSO, bilateral salpingo-oophorectomy; NA, not available; NED, no evidence of disease; DOD, died of disease.
attaches to other tissues, invades, and damages them. There are numerous reported cases of malignancy arising from endometriotic deposits and substantial histologic evidence that endometriosis is associated with endometrioid carcinoma and clear cell carcinoma of the ovary\(^{(11)}\). Atypical endometriosis possesses a precancerous potential, which is considered as premalignant changes of endometriosis characterized by cytologic atypia and architecture proliferation\(^{(1)}\). Our case showed a gradual transformation of benign endometriosis to atypical endometriosis and to malignant tumor, which satisfied Sampson and Scott diagnostic criteria\(^{(4,5)}\).

Although complete excision of the involved abdominal wall is the appropriate treatment for our case, the patient had recurrence soon after the operation. The pathology of the lesion also appears to be important to determine its course. The other most difficult aspect of wide excision is how to repair the defect of abdominal wall. Small fascia defect can be repaired with mesh effectively. For a big abdominal wall defect including the whole thickness of abdominal wall, skin graft and mesh repair might cause necrosis of grafted skin due to insufficient blood supply. Therefore, the use of autologous skin and muscle flap with its own blood supply to repair the defect is most appropriate. The use of local radiotherapy is limited because it might lead to severe skin reaction or even cause skin necrosis. The significance of chemotherapy for the management of a sarcoma remains controversial and it needs further evaluation and study. To our knowledge, this is the first case of carcinosarcoma arising from atypical endometriosis in a CS and is the first report on using autologous skin and muscle graft to repair the abdominal wall defect after excision. This case also highlights the potential for malignant change when recurrent scar endometriosis occurred after complete excision and ovarian expiration.

References


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