Case Report

Leiomyosarcoma of the Ovary Mimicking Gastrointestinal Stromal Tumor Originating from Small Bowel: A Case Report and Literature Review

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Abstract

Primary ovarian leiomyosarcoma is an extremely rare mesenchymal tumor that comprises less than 0.1% of all ovarian malignancies. Heterogeneous solid features on imaging and nonspecific bowel symptoms attributed to its close proximity to the gastrointestinal tract often cause confusion with a possible gastrointestinal tumor. We report a case of leiomyosarcoma of the ovary mimicking gastrointestinal stromal tumor from small bowel. Although rare, the possibility of primary ovarian leiomyosarcoma should be considered when the nature and location of a pelvic mass overlap with those of a gastrointestinal tract tumor.

Keywords: Leiomyosarcoma; Ovary; Gastrointestinal stromal tumor

Introduction

Leiomyosarcoma arising from the ovary is very rare, comprising less than 0.1% of all ovarian malignancies [1]. It usually affects postmenopausal women, with a few exceptions of cases in young women [2,3]. Because of its low prevalence and heterogeneous features on imaging, it is usually not diagnosed until pathologic examination following surgery. Vague digestive symptoms, such as lower abdominal discomfort, indigestion, and nausea, are also frequently encountered with tumors originating from the intestinal tract. These factors often lead to diagnostic confusion and therapeutic delay. However, not all ovarian leiomyosarcoma presents digestive symptoms, of which mainly presents urinary symptoms, or vaginal bleeding. Furthermore, there are no principal management recommendations for ovarian leiomyosarcoma due to absence of prospective studies [4]. Here, we present a rare case of primary ovarian leiomyosarcoma that was initially considered to be a gastrointestinal stromal tumor (GIST) originating from the small bowel.

We searched previous cases of primary ovarian leiomyosarcoma using PubMed system from 1965 to 2015 with the terms “primary ovarian leiomyosarcoma”. Then we analyzed and summarized the individual cases compared to our case.

Case Presentation

A 67-year-old, postmenopausal, multiparous woman presented to the gynecology clinic at Guro Hospital, College of Medicine, Korea University, with the complaint of a pelvic mass. Before visiting our hospital, she had undergone routine gynecologic checkup in a private clinic. At that time, transvaginal ultrasonography examination revealed an approximately 8-cm mixed echogenic solid mass of the right adnexa, which strongly suggested the presence of a malignant tumor (Figure 1). However, serum carbohydrate antigen (CA) 125 (17.5 U/mL), CA 19-9 (27.95 U/mL), carcinoembryonic antigen (CEA) (3.95 ng/mL), alpha fetoprotein (AFP) (3.05 ng/mL), and beta-human chorionic gonadotropin (beta-hCG) (0.26 mIU/mL) levels were not elevated. Pelvic magnetic resonance imaging (MRI) revealed an approximately 8-cm, lobulated, well-enhancing solid mass with a cystic portion in the pelvic cavity (Figure 2). The mass showed apparent diffusion restriction on diffusion-weighted MRI image. It had

![Figure 1: Transvaginal ultrasound showing mixed echogenic, solid mass measuring 8.9 × 7.3 cm.](image)

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an indistinguishable border with the distal ileum, but a fat plane was preserved between the mass and the sigmoid colon, uterus, and urinary bladder. Based on these findings, MRI suggested a mesenchymal tumor originating from the distal ileum, such as a GIST. However, a fibrotic tumor of right ovarian origin, such as a fibroma or fibrothecoma, was still possible. 18F-fluorodeoxyglucose (FDG) positron emission tomography (PET)/computed tomography (CT) scan also suggested a malignant mass. However, no lymph node enlargement or distant metastases were seen on imaging studies. Endoscopic examination of the stomach and colon showed no evidence of metastasis. Because MRI findings indicated GIST was likely, the patient was transferred to the general surgery department.

Explorative laparotomy with midline incision was performed. The surgical team found that the pelvic mass originated from the right ovary and adhered to the right pelvic wall and ureter. A team in the Department of Obstetrics and Gynecology took over patient care at that point and continued to perform the surgery. An 8-cm, multi-lobulated, irregular mass was visible in the right ovary (Figure 3). The mass was well-encapsulated, but some necrotic portion was also present. The right ureter was completely dissected from the right common iliac artery to the level of right uterine artery to separate the mass from the right pelvic wall. After adhesiolysis, right salpingo-oophorectomy was performed and the specimen was sent to the Department of Pathology for frozen examination. The result was a benign tumor, such as a fibroma. The uterus, left ovary, and left tube were normal on gross appearance. Total hysterectomy and left salpingo-oophorectomy was then performed. The operation was uneventful.

Histopathological analysis of the full specimen revealed a leiomyosarcoma, 9 × 7 × 5.6 cm³ in size, originating from the right ovary. The mitotic count was 12 per 10 high-power fields, and an area of necrosis was identified. The tumor cells had characteristically elongated, pleomorphic, and blunt-ended nuclei, and eosinophilic to pale cytoplasm (Figure 4). On immunohistochemical staining, tumor cells were positive for smooth muscle actin and desmin, but negative for calretinin, inhibin-alpha, and cluster of differentiation (CD) 34, consistent with a diagnosis of leiomyosarcoma. The uterus and left adnexa were free of metastasis. Postoperatively, the patient has been followed up for 3 months without any adjuvant treatment.

After excluding cases of leiomyosarcomas originating from ovarian vein, six cases of primary ovarian leiomyosarcomas were found by a PubMed search system (Table 1) [2,5-9]. Among them, only 2 cases showed elevated CA 125 level. Postoperative adjuvant chemotherapy was performed in 2 cases. Immunohistochemical studies revealed that smooth muscle actin was positive in most of cases, whereas inhibin was negative. During follow up, only 1 patient expired and 4 had no evidence of disease.

Before writing this case report, we obtained informed consent for the publication from the patient.

**Discussion**

Leiomyosarcomas account for 10–20% of all soft tissue sarcomas [10]. They most frequently arise from the uterus, gastrointestinal tract, and retroperitoneal region [11]. Those originating from the ovary are exceptionally rare, comprising less than 1% of ovarian sarcomas [1]. Other types of ovarian sarcomas include carcinosarcoma, rhabdomyosarcoma, stromal cell sarcoma, and fibrosarcoma [12].

Primary ovarian leiomyosarcomas can be confused with leiomyosarcomas of extrabdominal origin. For example, pedunculated uterine leiomyomas can attach to the ovary from which they draw their blood supply. In addition, leiomyosarcomas originating from broad ligaments, ovarian veins, and the gastrointestinal tract should be distinguished from primary ovarian leiomyosarcomas [5]. Identification of normal ovarian components in tumor tissue
categorizes a tumor as a primary ovarian leiomyosarcoma. Low disease prevalence and non-specific radiological findings often hinder accurate diagnosis of this tumor preoperatively.

Interestingly, most leiomyosarcomas of ovaries were provisionally misdiagnosed of another disease due to its rarity and diagnostic difficulty, which is consistent with our case. Bouie et al. initially considered the ovarian leiomyosarcoma as a fibrothecoma [2]. Divya et al. made a radiologic diagnosis of broad ligament fibroid because the mass was located ischial fossa displacing the rectum [8]. In addition, Vijaya et al. supposed the mass to be an ovarian leiomyoma on the basis of ultrasonography and MRI [9].

In this case, MRI initially suggested the tumor mass was a mesenchymal tumor originating from the distal ileum, such as a GIST. GIST is the most common mesenchymal tumor of the gastrointestinal tract [13]. It can originate at any site along the intestinal tract, including the stomach, jejunum, ileum, duodenum, rectum, colon, and esophagus. Radiologic features of GISTs vary depending on their size and anatomic location. Small (<=5 cm) GISTs are round and have strong homogeneous arterial and persistent enhancement. In contrast, larger GISTs (>5 cm) are frequently lobulated and have mild, heterogeneous, gradual enhancement and heterogeneity with intratumoral hemorrhage, necrosis, or cystic change [14]. Unfortunately, these findings are also commonly encountered in patients with ovarian leiomyosarcoma, as in this case. For these reasons, preoperative radiological differentiation between ovarian leiomyosarcoma and GIST is difficult. A large tumor mass abutting the gastrointestinal tract, as in this case, makes the diagnosis more challenging. In this case, preoperative endoscopic evaluation of the stomach and colon did not identify any mass lesion. However, these negative findings cannot completely rule out the presence of GIST because these two tests do not cover the small intestine. If barium swallow studies such as small bowel series are performed, the possibility of small bowel GIST could easily be eliminated.

Another important point in this case was subsequent management according to intraoperative frozen examination. The result indicated a benign mass, such as a fibroma. Although the uterus, left ovary, and left tube were normal on gross appearance, total hysterectomy and left salpingo-oophorectomy were performed due to the possibility of malignant findings on final pathologic examination. At present, the main treatment modality for ovarian leiomyosarcoma is surgery. This ranges from fertility preserving surgery to debulking surgery including total hysterectomy, bilateral salpingo-oophorectomy, and excision of any suspicious masses. In this case, the patient was postmenopausal and had no need to preserve fertility, so debulking surgery was performed.

Given the very poor prognosis of leiomyosarcoma, misdiagnosis may have a detrimental effect on patient outcome. In this case, all preoperative evaluations, including serum tumor markers, identified GIST as the most probable diagnosis. Unfortunately, no specific tumor markers have exhibited clinical utility for detecting leiomyosarcomas. Pathologically, leiomyosarcomas can be distinguished from leiomyomas by the following features: larger tumor cells, cytologic atypia, increased mitotic activity, area of necrosis, and nuclear pleomorphism, consistent with our case [3,15]. Immunohistochemical markers including desmin, vimentin, and smooth muscle actin are used to diagnose leiomyosarcoma, whereas CA125 and CD34 are well-known specific markers for GIST [16].

Given the rarity of this disease, the benefit of adjuvant chemotherapy or radiation therapy has not been established. Vijaya et al. reported that chemotherapy with vincristine, epirubicin, and cyclosphosphamide was beneficial to prolong progression-free survival [9]. Hensley et al. used a combined gemcitabine and docetaxel regimen, frequently used for uterine leiomyosarcoma, and demonstrated disease-free interval for 22 months [17]. Radiotherapy has been shown as temporarily effective for local control, but it failed to prevent distant metastasis [18].

In this study, we presented a case of primary ovarian leiomyosarcoma initially confused with GIST originating from the distal ileum. This case report suggests an important lesson. When a pelvic mass is suggestive of GIST on radiological evaluation but could still be ovarian cancer based on anatomic location or imaging features, ovarian sarcoma should be suspected. Another way to differentiate between these entities is to perform a barium swallow study to examine the entire intestinal tract. Appropriate surgical treatment should not be delayed solely on the basis of normal tumor marker levels. Because of its dismal prognosis, early diagnosis and prompt surgical removal are crucial for patients with primary ovarian leiomyosarcoma.

Table 1: Summary of previous cases of primary ovarian leiomyosarcoma.

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age (yr)</th>
<th>Size (cm)</th>
<th>Tumor marker</th>
<th>Type of surgery</th>
<th>Adjuvant treatment</th>
<th>Positive</th>
<th>Weakly positive</th>
<th>Negative</th>
<th>Follow-up (mo)</th>
<th>Current status</th>
</tr>
</thead>
<tbody>
<tr>
<td>Monk et al. [5]</td>
<td>1993</td>
<td>12</td>
<td>17 x 13.5 x 10.5</td>
<td>AFP (-), beta-hCG (-), CEA (-)</td>
<td>BSO</td>
<td>No further treatment</td>
<td>SMA</td>
<td>Desmin</td>
<td>ND</td>
<td>20</td>
<td>NED</td>
</tr>
<tr>
<td>Seracchioli et al. [8]</td>
<td>2003</td>
<td>20</td>
<td>8 x 8</td>
<td>Negative, but type of marker was not described</td>
<td>LSO</td>
<td>No further treatment</td>
<td>SMA, p53</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>NED</td>
</tr>
<tr>
<td>Bouie et al. [7]</td>
<td>2005</td>
<td>42</td>
<td>18 x 13 x 5</td>
<td>ND</td>
<td>TAH LSO</td>
<td>No further treatment</td>
<td>SMA, Vimentin, Ki-67, Desmin</td>
<td>AE1/AE3, p53</td>
<td>Inhibin, HMBr45, S100</td>
<td>3</td>
<td>NED</td>
</tr>
<tr>
<td>Dai et al. [8]</td>
<td>2011</td>
<td>42</td>
<td>12</td>
<td>CA125 (39.4)</td>
<td>Staging surgery</td>
<td>cisplatin, etoposide, bleomycin</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>5</td>
<td>Expired</td>
</tr>
<tr>
<td>Divya et al. [9]</td>
<td>2014</td>
<td>26</td>
<td>9 x 6 x 3</td>
<td>ND</td>
<td>RSO</td>
<td>No further treatment</td>
<td>SMA, Vimentin</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>NED</td>
</tr>
<tr>
<td>Vijaya et al. [10]</td>
<td>2015</td>
<td>30</td>
<td>14.7 x 14.6 x 10.8</td>
<td>CA125 (69.9)</td>
<td>RSO</td>
<td>Vincristine, epirubicin, cyclophosphamide</td>
<td>SMA, Vimentin, Desmin</td>
<td>ND</td>
<td>Inhibin</td>
<td>6</td>
<td>NED</td>
</tr>
<tr>
<td>This case</td>
<td>2015</td>
<td>67</td>
<td>9 x 7 x 5.6</td>
<td>CA125 (+), CA 19-9 (+), CEA (+), AFP (-), beta-hCG (-)</td>
<td>TAH BSO</td>
<td>No further treatment</td>
<td>SMA, Desmin</td>
<td>ND</td>
<td>Inhibin, CA125, CD34</td>
<td>3</td>
<td>NED</td>
</tr>
</tbody>
</table>

AFP: alpha fetoprotein; hCG: human chorionic gonadotropin; CEA: carcinoembryonic antigen; ND: not described; CA: Carbohydrate antigen; BSO: bilateral salpingo-oophorectomy; LSO: left salpingo-oophorectomy; TAH: total abdominal hysterectomy; RSO: right salpingo-oophorectomy; SMA: smooth muscle actin; HMB: human melanoma black; CD: Cluster of differentiation; NED: no evidence of disease

References