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Primary ovarian leiomyosarcoma

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Summary

Background: Primary ovarian leiomyosarcoma is an extremely rare subtype of ovarian sarcomas. It most commonly occurs in postmenopausal women and has unfavorable prognosis. *Case:* The patient, a 58-year-old postmenopausal woman, presented with a complaint of abdominal pain. Preoperative examination revealed an intraabdominal mass 25 x 17 x 14 cm in the right adnexa. She underwent bilateral salpingo-oophorectomy, total omentectomy, appendectomy and bilateral pelvic lymphadenectomy. The histopathology revealed leiomyosarcoma of the right ovary Stage Ia. She did not receive any postoperative adjuvant therapy. Follow-up 21 months after initial surgery, showed no evidence of recurrence. *Conclusion:* Additional studies are needed to understand more about the nature, clinical behavior and treatment of this very rare tumor.

Key words: Primary ovarian leiomyosarcoma; Treatment; Surgery; Radiotherapy; Chemotherapy; Prognosis.

Introduction

Primary ovarian sarcoma is a very rare tumor accounting for less than 3% of all ovarian malignancies [1]. The most common histologic subtypes are carcinosarcoma, endometrial stromal sarcoma, fibrosarcoma and rhabdomyosarcoma [2-4]. Leiomyosarcoma is an extremely rare subtype of ovarian sarcomas [5].

Until now, about 64 cases of primary ovarian leiomyosarcoma (POLMS) have been reported in the English literature [6]. POLMS most commonly occurs in postmenopausal women and has unfavorable prognosis [2, 5-7]. We present a case of POLMS and review the literature.

Case Report

The patient, a 58-year-old, gravida 1, para 1 postmenopausal woman presented with a complaint of abdominal pain. She had a history of hysterectomy for uterine leiomyomas without salpingo-oophorectomy 15 years before. Her family history revealed no evidence of cancer among the first-degree relatives.

On gynecologic examination there was a palpable pelvic mass. There were no palpable inguinal lymph nodes and the rest of pelvic examination was normal.

Preoperative computed tomography (CT) of the abdomen and pelvis, and abdominal ultrasound (US) revealed an intraabdominal mass 25 x 17 x 14 cm in the right adnexa. Preoperative CT of the chest, chest X-ray, colonoscopy and urethrocystoscopy were normal. Preoperative intravenous pyelography (IVP) revealed bilateral distention of the ureter and hydronephrosis. Preoperative CA-125 was elevated to 63.4 U/ml.

On exploratory laparotomy, the right ovary was markedly distended, measuring 25 x 17 cm. Frozen section showed malignancy and the patient underwent bilateral salpingo-oophorectomy, total omentectomy, appendectomy and bilateral pelvic lymphadenectomy.

Histopathology revealed leiomyosarcoma of the right ovary (Figures 1, 2). Tumor cells had high mitotic activity (11 mitotic figures per 10 high-power fields). Tumor was limited to the right ovary without penetrating the serosal surface. The peritoneal washing smear was negative for malignant cells. Histologic diagnosis was confirmed by positive immunostaining. Tumor cells were positive for smooth muscle actin (SMA), vimentin, desmin and ki-67, weakly positive for S-100 protein and negative for epithelial membrane antigen (EMA), cytokeratin, inhibin, CD10 and CD99. The final diagnosis was Stage Ia leiomyosarcoma of the right ovary.

The patient did not receive any postoperative adjuvant therapy. Follow-up 21 months after initial surgery with CT of the chest, abdomen and pelvis, abdominal US, chest X-ray, IVP, colonoscopy and urethrocystoscopy showed no evidence of recurrence.

Discussion

POLMS is a very rare tumor [5]. Until now, about 64 cases of POLMS have been reported in the English literature [6]. Among them, 41 were POLMS without heterologous elements and 23 were POLMS with heterologous elements [5-8].

The precise histogenesis of POLMS is still uncertain [5]. It probably originates from smooth muscle present in the walls of the blood vessels in the cortical stroma, in the corpus luteum, in the ovarian ligaments at their point of attachment to the ovary, in remnants of Wolffian ducts and in totipotential cells of ovarian mesenchyme [5, 9-11]. Malignant transformation of an ovarian leiomyoma and migration of a uterine leiomyoma are very rare mechanisms of histogenesis [12, 13].

POLMS usually occurs in postmenopausal women, although sometimes younger women may be affected [2, 7, 14]. It has been reported in patients between 12 and 84 years of age (mean age 52.6 years) [7, 8]. It is usually unilateral and may reach more than 10 cm in diameter [6, 7, 11, 12]. In our case, the patient was 58 years old and POLMS was unilateral.

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Figure 1. — Primary ovarian leiomyosarcoma (HE stain x100).

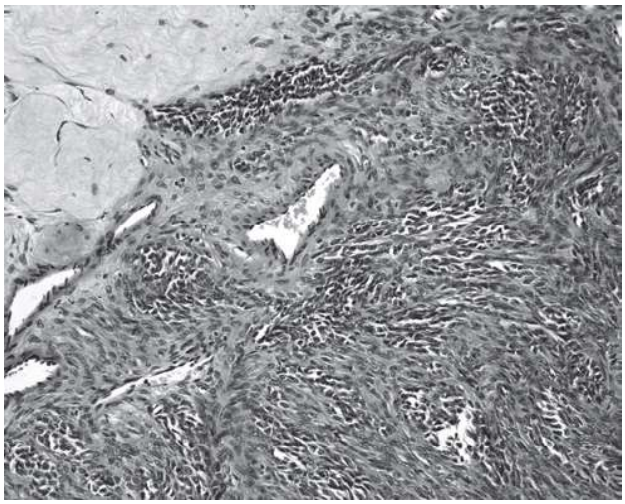

Figure 2. — Primary ovarian leiomyosarcoma (HE stain x200).

Most patients with POLMS usually have nonspecific symptoms and signs such as pelvic pain and abdominal bloating [6, 13]. Also they have symptoms of pressure on the bladder and bowel [13]. Our patient had abdominal pain with no other symptoms and signs.

POLMS is typically present as a solitary, large, lobular, soft, fleshy, solid mass with hemorrhage and cystic degeneration [6, 12]. The diagnostic criteria for POLMS are hypercellularity, nuclear atypia, pleomorphism, coagulative necrosis and high mitotic activity (> 5 mitotic figures per 10 high-power fields) [6, 9, 12]. According to these criteria, our case is POLMS, as it fulfills all of them.

Immunohistochemical staining for POLMS is generally positive for muscle specific actin, SMA, desmin, vimentin, p53 and proliferation markers and negative for cytokeratins and S-100 [2, 8, 11, 12, 15]. Mitotic activity and stage have a direct correlation with the malignant potential and aggressiveness of POLMS [5]. In our patient, tumor cells were positive for SMA, desmin, vimentin and ki-67, weakly positive for S-100 protein and negative for EMA, cytokeratin, inhibin, CD10 and CD99.

Due to the rarity of POLMS it is very difficult to determine the optimal therapy [5]. Surgery remains the mainstay of treatment for POLMS and complete resection should be attempted whenever possible [7, 13]. Initial debulking surgery usually consists of total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and exstirpation of all resectable tumor masses within the pelvis and abdomen [5, 7, 16, 17]. In our case, the patient underwent bilateral salpingo-oophorectomy, total omentectomy, appendectomy and bilateral pelvic lymphadenectomy.

Postoperative radiotherapy is used for local disease control [13]. Postoperative chemotherapy is used for prevention of distant metastases [7, 13]. Various chemotherapeutic regimens including cisplatin have been applied [5, 7]. However, the role of postoperative radiotherapy or chemotherapy remains controversial, especially in patients with disease confined to the ovary [2, 5, 7, 13, 16, 18]. Postoperative adjuvant therapies should be attempted for unresectable residual disease because those patients have a dismal prognosis [7]. Our patient was Stage Ia, so she did not receive any postoperative adjuvant therapy.

POLMS most commonly recurs in the abdomen and pelvis [8]. Less frequently recurrence is in the lung, bone, liver, mediastinum and brain [8].

The prognosis of POLMS is generally unfavorable, depending on mitotic activity and stage at diagnosis [2, 5]. Most reported cases recurred within one year and patients died within two years after initial diagnosis [7]. Patients die with extended local disease and multiple distant metastases [13]. Our patient was Stage Ia and, 21 months after initial surgery, she is well with no evidence of recurrence.

Conclusion

POLMS is a very rare tumor and has unfavorable prognosis. Additional studies are needed to understand more about the nature, clinical behavior and treatment of this tumor.

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