Primary ovarian leiomyosarcoma: A case report

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Summary

Primary ovarian leiomyosarcomas are extremely rare tumors that comprise less than 0.1% of all ovarian malignancies. We present a case of 62-year-old postmenopausal woman with a slightly enlarged right ovary and a Color Doppler sonography resistance index (RI) measuring 0.54. The patient, after being managed with surgery alone, is alive after 14 months without any evidence of disease. This is an unusual case in that primary ovarian leiomyosarcoma was diagnosed in the setting of a slightly enlarged irregular postmenopausal ovary with a concomitant intermediate RI value on color flow Doppler evaluation. A high index of suspicion may help prevent delay in the diagnosis of this rare neoplasm.

Key words: Primary ovarian leiomyosarcoma; Color flow Doppler.

Introduction

Ovarian sarcomas are rare neoplasms that comprise less than 1% of all ovarian malignant tumors. As with all sarcomas, ovarian sarcomas may be low or high grade and may contain elements of purely mullerian origin (homologous) or may have nonovarian elements (heterologous).

Primary ovarian leiomyosarcomas which are high-grade sarcomas arise from smooth muscle elements normally found in the hilus of the ovary. Overall survival with ovarian sarcomas has been poor, particularly when metastatic disease is found at the time of surgery. Most patients die of disease within two years of diagnosis although long-term survivors have been reported in every series [1].

We present an unusual case of primary ovarian leiomyosarcoma arising in a slightly enlarged postmenopausal ovary.

Case Report

A 62-year-old, gravida 7, para 5, postmenopausal woman presented to the Department of Obstetrics and Gynecology at Baskent University Faculty of Medicine, with the chief complaint of nocturia and urge incontinence. The patient was in good medical condition, did not complain of pain or weight loss and anamnesis was unremarkable. As for her medical history, she was taking anti-hypertensive medication but she had never been on hormone replacement therapy. On pelvic examination she was found to have mild systocele. Transvaginal ultrasonography revealed a 15-mm fundal myoma on an atrophic uterus with a lobulated right ovary. It measured 33 x 30 x 24 mm in size and was larger than the atrophic left ovary. With the suspicion of detecting a right ovarian solid tumor, color Doppler sonography was performed. The resistance index (RI) and pulsatility index (PI) values were found to be 0.54 and 1.40, respectively. Serum tumor markers CA-125, alpha-fetoprotein, lactate dehydrogenase and β-hCG were normal. Based on these findings, a diagnostic laparoscopy was undertaken where no further findings consistent with malignancy were noted. Bilateral salpingo-oophorectomy was performed and both ovaries were sent for frozen section analysis. The left ovary was 2.50 x 1.0 x 0.7 cm in size with corpus albicans revealed on the cut surface. The right ovary was 3.5 x 3.0 x 2.0 cm in size with irregular borders and its cut surface revealed yellowish cream-colored solid areas. Frozen section analysis was reported as fibrothecoma. Five days after laparoscopy, as the final pathology depicted right ovarian leiomyosarcoma, the diagnosis was discussed with the patient and a staging laparotomy was performed. There were no signs of metastatic disease in the abdomen. Peritoneal cytology and multiple biopsies were taken and total abdominal hysterectomy, bilateral pelvic and paraaortic lymph node sampling, omentectomy, and appendectomy were performed. No tumoral metastasis was found. In cross sections of the right ovary, a tumor consisting of smooth muscle cells forming bundles was detected (Figure 1). Cells in some focal areas were noted to have mild degrees of pleomorphism, swelling, and epithelioid features. Overall the tumor showed 5-12 mitoses per 10 high power fields (HPF) with up to 22 mitotic figures in selected 10 HPFs. There was no necrosis whereas nuclear atypia was present (Figure 2). Immunohistochemical studies revealed diffuse and strongly positive smooth muscle actin and vimentin while desmin was observed to be moderately positive in focal areas, and there was no expression of progesterone receptor. An immunohistochemical study for proliferating cell nuclear antigen (PCNA) was done to evaluate the proliferative index of the tumor and 90-95% of tumor cells were positive. The uterus was 5.0 x 4.0 x 3.5 cm in size in which a subserous myoma measuring 0.5 x 0.6 x 0.5 cm was present. The postoperative recovery was unremarkable. No further treatment was given and the patient was alive without any evidence of disease 20 months after surgery.

Discussion

Primary ovarian leiomyosarcomas are rare tumors that comprise less than 0.1% of all ovarian malignancies [2].

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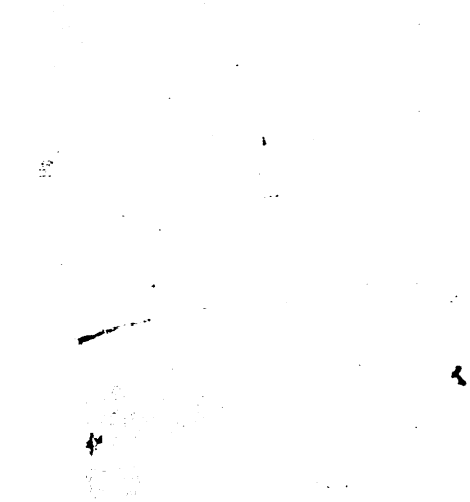

Figure 1. — Tumor consisting of bundles of smooth muscle cells next to the ovarian stroma (H&E x 40).

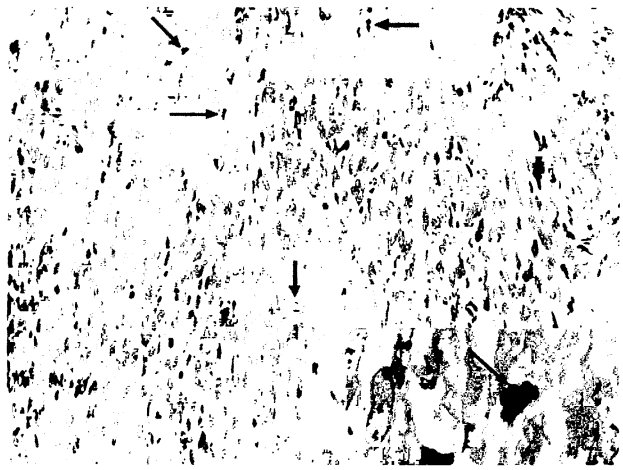

Figure 2. — Ovarian tumor showing pleomorphism, hyperchromatic nuclei and multiple mitotic figures (arrows) (H&E x 200). Inset: An abnormal mitotic figure in tumor cells (H&E x 400).

To our knowledge there are only 35 reported cases in the literature. The usual presentation is a postmenopausal woman with a large pelvic mass. On the contrary, in the case presented herein the patient did not have a pelvic mass, but only a slightly enlarged right ovary with irregular contours detected at transvaginal ultrasonography. Color Doppler sonography indicated a RI of 0.54, which is an intermediate value between benign and malignant masses. High impedance values (PI > 1.5, RI > 0.6) may be consistent with a probably benign histology, whereas low impedance values (PI < 1.0, RI < 0.4) may be more likely consistent with malignant histology [3].

The postmenopausal gonad atrophies to a size of 1.5 x 1.0 x 0.5 cm on the average and at that size it should not be palpable on pelvic examination. Only 10% of patients with a palpable postmenopausal ovary who are subjected to oophorectomy are found to have a malignant ovarian neoplasm. Lerner *et al.* accurately predicted a benign outcome in 247 of 248 patients with a palpable ovary using color flow Doppler analysis [4].

The origin of primary ovarian leiomyosarcomas is thought to be similar to leiomyomas, which probably originate from smooth muscle cells present in the walls of blood vessels in the cortical stroma, in the corpus luteum, and in the ovarian ligaments at their points of attachment to the ovary [5]. Leiomyosarcomas can also originate from the smooth muscle component of teratomas, and totipotency of the ovarian mesenchyme may be one of the underlying mechanisms [6].

The histopathologic features of leiomyomas may occasionally deviate from the norm, hence several subtypes have been described [7]. Among these subtypes, cellular leiomyomas, mitotically active leiomyomas and atypical leiomyomas are important in the differential diagnosis of leiomyosarcomas. Cellular leiomyomas lack significant cytologic atypia, and mitotic activity in this lesion usually does not exceed 4 normal mitoses per 10 HPF [8]. In contrast to cellular leiomyomas, leiomyosarcomas, even those with absent-to-minimal cytologic atypia, exhibit a mitotic count usually greater than 10 mitoses per 10 HPF. Mitotically active leiomyomas are otherwise normal leiomyomas which are usually associated with more than 5 and a maximum of 20 normal mitoses per 10 HPF [9]. Atypical leiomyomas are characterized by moderate-to-severe cytologic atypia and 0 to 4 mitoses per 10 HPF. Tumor cell necrosis typically is absent in these lesions [10]. Differentiating an atypical leiomyoma from a leiomyosarcoma may be difficult, hence extensive sampling of the tumor is prudent.

Due to the rarity of reported cases, there are no established diagnostic criteria and therapeutic strategies for ovarian leiomyosarcomas, and the prognosis is also uncertain. Criteria applied for the diagnosis of uterine leiomyosarcomas are also currently acceptable for ovarian leiomyosarcomas [11, 12]. According to studies of Bell *et al.* on uterine smooth muscle tumors, malignant behavior is almost always limited to tumors with any two of the sentinel histopathologic features: coagulative tumor cell necrosis, diffuse moderate to severe cytologic atypia, and a mitotic index greater than 10 [9]. Immunohistochemistry and electron microscopy also aid in the diagnosis of this rare tumor. Desmin and smooth muscle actin are expressed in primary ovarian leiomyosarcoma [11].

There is no conclusive evidence that adjuvant chemotherapy or radiation offers any survival advantage for primary ovarian leiomyosarcoma. Cortes *et al.* and Monk *et al.*, believe that surgery remains the mainstay of treatment and any adjuvant therapies are of unproven benefit [2, 13].

This is an unusual case in that primary ovarian leiomyosarcoma was diagnosed in the setting of a slightly enlarged irregular postmenopausal ovary with a concomitant intermediate RI value on color flow Doppler evaluation. A high index of suspicion may help prevent delay in the diagnosis of this rare neoplasm.

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