



Oncology

A Case of Retroperitoneal Metastases That Occur 14 Years After Surgery



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ABSTRACT

Endometrial Stromal Sarcomas are rare malignant tumours of the uterus. We report the case of incidental finding of Low-Grade Endometrial Stromal Sarcoma (LGESS) that metastasized to the retroperitoneum 14 years after the original surgery in a 72-year-old woman. The patient underwent a laparotomy and excision of all tumour nodules. Considering the common recurrence of and slow growing nature of LGESS, appropriate treatment options like surgical excision and life-long follow up should be considered. © 2016 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Introduction

Malignancy of the uterine corpus is the fourth most common cancer in women living in developed countries worldwide. Endometrial Stromal Sarcoma (ESS) is a rare neoplasm comprising only 0.2% of all uterine malignancies and 15–26% of primary uterine sarcomas with less than 700 cases diagnosed annually.¹ ESS was formerly classified into two distinct subtypes, low-grade and high-grade, based on differences in morphological atypia and proliferative activity. The World Health Organization (WHO) re-classified these entities into three groups: benign endometrial stromal nodule (ESN), low-grade endometrial stromal sarcoma (LGESS), and undifferentiated endometrial sarcoma (UES).² LGESS is a slow growing tumor with good prognosis and a 50% recurrence following treatment. Common metastatic sites for LGESS are the vagina, pelvis, and peritoneal cavity although distant metastasis to sigmoid colon, lung, liver, brain, has also been reported.³ We present a case of low-grade ESS that recurred 14 years following the original surgery with metastasis in the retroperitoneal.

Case presentations

A 72-year-old woman presented with dyspnea and bilateral peripheral edema. She had undergone total hysterectomy for low-grade

endometrial stromal sarcoma (LGESS) tumor fourteen years prior, which was confirmed histologically to be LGESS. There were no significant past medical history or history of familial disease. Physical examination showed palpable abdominal mass at the Right Lower Quadrant and Left Lower Quadrant. The following tumor markers (AFP, CEA, Cancer Antigen 125, and BhCG) are within reference limits. Yet, normal results do not exclude neoplasia. CT scan of the abdomen and pelvis revealed presence of several retroperitoneal masses surrounding the abdominal aorta and vena cava (Fig. 1). Percutaneous biopsy of the lesion showed diffuse proliferation of spindle-shaped cells with little nuclear atypia. Morphologically, the tumor resembles the tumor from the 1993 histopathology specimen.

The patient underwent a laparotomy and excision of all tumor nodules. Seven nodular masses were excised weighing altogether 1042 g (Fig. 2). The largest nodule measured 160 × 100 × 80 mm in dimension (Fig. 3) and the smallest was 20 × 20 × 12 mm. The largest nodule impinges upon the right ureter, right kidney and iliac vessels. The external surfaces of all these nodules appear lobulated, multinodular, and covered by a serosal surface that formed a capsule. This enabled easy excision during surgery.

The patient's postoperative course was uneventful. She was free of symptoms 2 weeks following abdominal surgery.

Discussion

Clinical characteristics of low-grade ESS (LGESS) include a slow growth and indolent disease course with a tendency for

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late recurrence. A study by Piver et al reported that the intervals before recurrence varied from 3 months to 23 years, with a median interval of 3 years. 50% of patients with low-grade ESS develop recurrences or metastases in the vagina, pelvis, and peritoneal cavity although distant metastasis has been reported.⁴

Although outdated, ESS was formerly classified into low-grade ESS and high-grade ESS based on differences in the cell's mitotic activity. More than 10 mitotic figures for high-grade ESS and less than 10 mitotic figures per 10 HPF for low-grade ESS. Microscopic examination of the tumor showed that it has mitotic count less than

10 in mitosis per high power fields (HPF) – thus classifying the tumor as low-grade ESS rather than high-grade ESS.

Surgical excision is currently the only therapeutic procedure for LGESS. Standard treatment for its recurrent disease such as radiotherapy and chemotherapy has not been established to be effective. Immunoreactivity for estrogen receptor and progesterone receptor is not regularly assessed in LGESS. Based on several clinical studies, Mansi et al recommended that progesterone therapy should be the treatment of first choice for relapsed LGESS because there was resolution or stabilization of recurrent or metastatic disease in more than 50% of patients

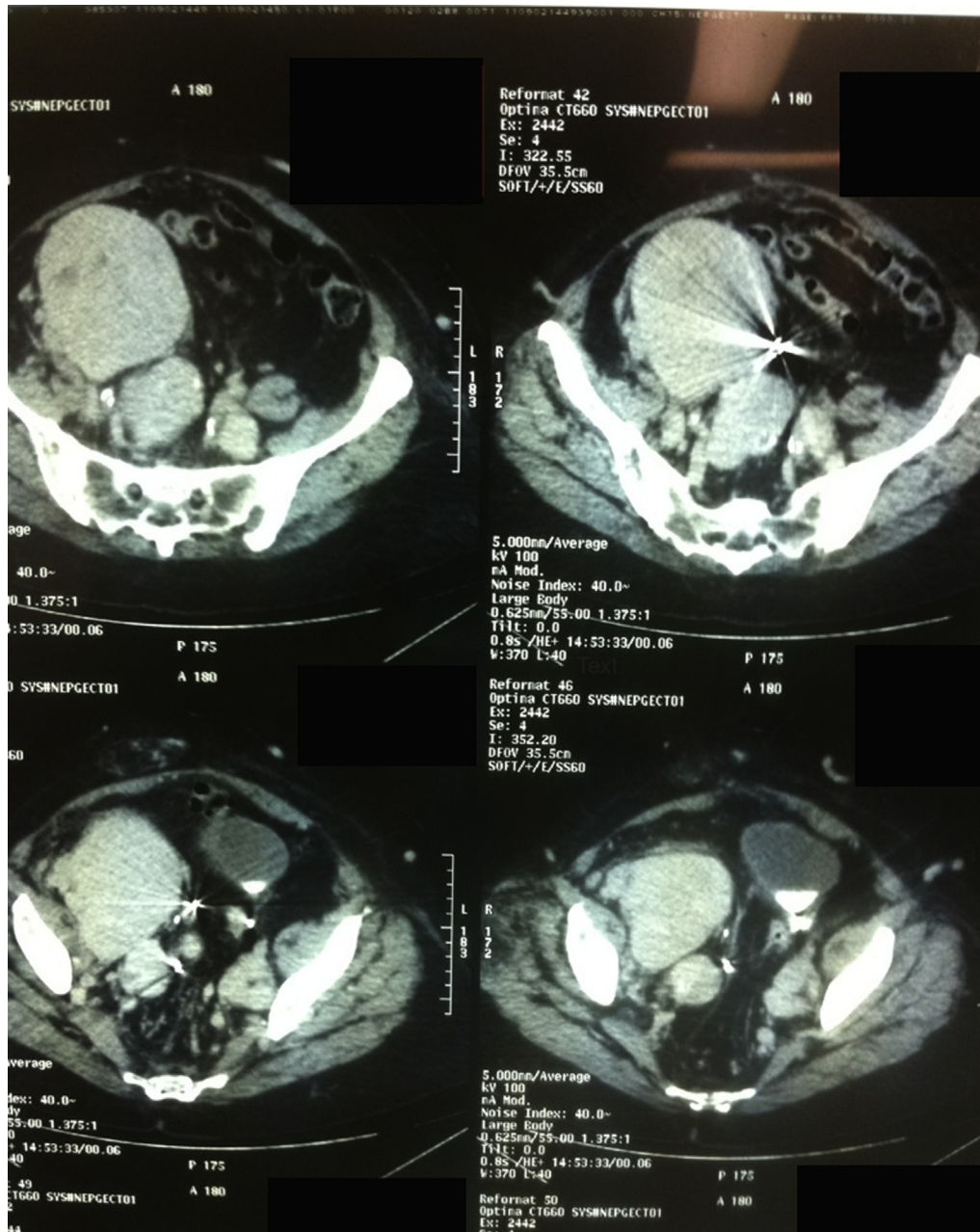


Figure 1. Abdominal & pelvic CT scans which revealed presence of several retroperitoneal masses surrounding the abdominal aorta and inferior vena cava.

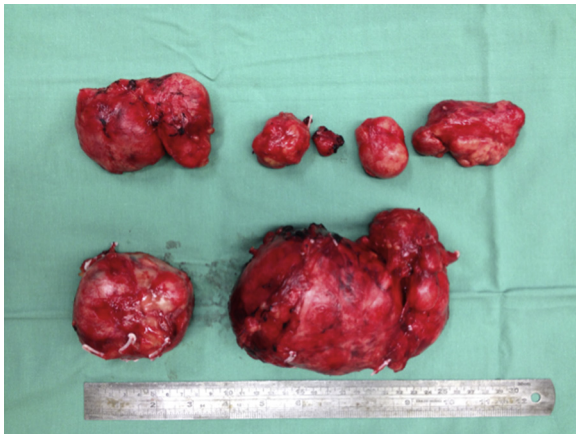


Figure 2. Seven nodular masses that were excised. They weighed 1042 g with the largest measuring 160 × 100 × 80 mm and the smallest measuring 20 × 20 × 12 mm.

treated with progestational agents.⁵ This suggestion is noteworthy since our tumor sample showed strong and diffuse staining against anti-estrogen and progesterone receptor antibodies.



Figure 3. The largest nodule excised. It impinges upon the right ureter, right kidney, and iliac vessels. Causing lower extremity edema.

Conclusion

Considering the slow growing nature and common recurrence of LGEES, 14 years' recurrences in this case, a life-long follow up and routine assessment for anti-estrogen and anti-progesterone should be integrated into clinical practice in LGEES management. Any sign of recurrence should be managed with surgical excision.

Ethical approval

For this type of study, no formal ethical approval is necessary.

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None.

Authors contribution

Masjensen Argadjendra wrote the first version of the manuscript, collected and analyzed the data and was involved in patient care.

Nariman Ahmadi was involved in patient care, analyzed the data and contributed to the manuscript.

Celi Varol was involved in patient care, analyzed the data, proposed the study concept and contributed to the manuscript.

Conflict of interest

The authors declare no conflict of interest.

Consent

Written informed consent was obtained from the patients for publication of this case report and accompanying images.

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