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Published online 2015 August 1. **Case Report**

Heart and Lung Metastases From Endometrial Stromal Sarcoma in a Forty-Two-Year-Old Woman

Behnam Shakerian [1]; Mohammad Hossein Mandegar [1]; Bahieh Moradi [2,*]; Farideh Roshanali [2]

[1]Department of Cardiothoracic Surgery, Shariati General Hospital, Tehran University of Medical Sciences, Tehran, IR Iran
[2]Department of Echocardiography, Day General Hospital, Tehran, IR Iran

*Corresponding author: Bahieh Moradi, Department of Echocardiography, Day General Hospital, P. O. Box: 1434873111, Tehran, IR Iran. Tel: +98-2184942875, Fax: +98-2166005214, E-mail: faraviolet@yahoo.com

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Introduction: Low-grade endometrial stromal sarcoma (LG-ESS) is a malignant intrauterine tumor that rarely presents with distant metastasis. Simultaneous lung and cardiac metastases from LG-ESS is also an extremely rare event.
Case Presentation: A 42-year-old woman presented with dyspnea and exercise intolerance. She had a history of hysterectomy and left salpingoophorectomy. She underwent second laparotomy as well as right oophorectomy after new finding of vaginal mass with histopathologic diagnosis of LG-ESS. Cardiac imaging techniques demonstrated tumoral process in the right atrium and ventricle, coronary sinus, and pulmonary outlet tract as well as multiple metastases in the lung fields. Successful complete surgical resection of the metastatic tumor in the right side of the heart and then radiotherapy were done. After 28 months, follow-up examination revealed no abnormality.
Conclusions: We describe the first documented case of isolated intracardiac and lung metastases of a LG-ESS without concurrent abdominal or caval metastasis.

Keywords: Sarcoma; Surgery; Heart Neoplasms

1. Introduction

Endometrial stromal tumors are uncommon tumors, which mostly develop in the uterus, and occasionally in the ovary and peritoneum (1). The endometrial stromal sarcoma (ESS) is a rare disease and represents 0.2% of all uterine neoplasm, with fewer than 450 new cases diagnosed annually in the United State (2, 3). The world health organization currently divides these tumors into four different subtypes based on clinical and pathologic features: endometrial stromal nodule (ESN), the malignant subtypes, low-grade ESS (LG-ESS) and high-grade endometrial stromal sarcoma (HG-ESS) as well as undifferentiated uterine sarcoma (UUS). The HG-ESS as well as UUS comprise smaller than 10% of uterine sarcomas and smaller than 1% of all primary malignant tumors of the uterus (1, 2).

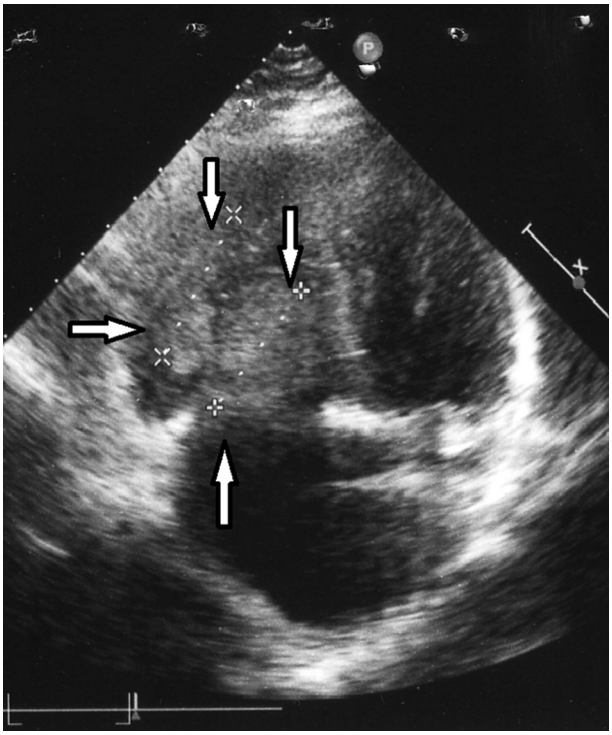
Simultaneous lung and isolated intracardiac metastasis from LG-ESS is an extremely rare event. We described a patient who successfully underwent complete surgical resection of metastatic tumors of the LG-ESS in the right side of the heart. It was the first documented case of isolated intracardiac and lung metastasis of ESS without concurrent abdominal or caval metastasis in English literature.

2. Case Presentation

A 42-year-old woman presented with dyspnea and reduced exercise tolerance, which had begun a month before admission. Her medical history included hysterectomy and left salpingoophorectomy for menorrhagia and a left ovarian mass five years ago. Histopathologic examination demonstrated multiple leiomyomas and fibrothecomas of the ovary. Four months later, she complained of spotting. Transvaginal ultrasonography showed a heterogeneous solid mass, about 28 mm × 45 mm, in the cuff of the vagina. She underwent laparotomy and excision of the mass as well as right oophorectomy. Histopathologic examination revealed a LG-ESS. She underwent 28 courses of radiotherapy.

On examination, the patient had dyspnea at rest, tachycardia, and normal blood pressure. She had mild to moderate lower limb edema. Transthoracic echocardiography showed a large multilobulated mass (4.5 cm × 3.5 cm) in the right ventricle (Figure 1). Computed tomographic (CT) angiography demonstrated tumoral process in the right atrium, coronary sinus, right ventricle, and pulmonary outlet tract as well as multiple metastases in both lung fields (Figure 2). Findings of abdominopelvic and brain CT scan were normal. Color Doppler ultrasonography of the vessels of both lower limbs revealed normal findings.

Figure 1. A Large Multilobulated Mass in the Right Ventricle



White arrows, apical 4-chamber view of transthoracic echocardiography.

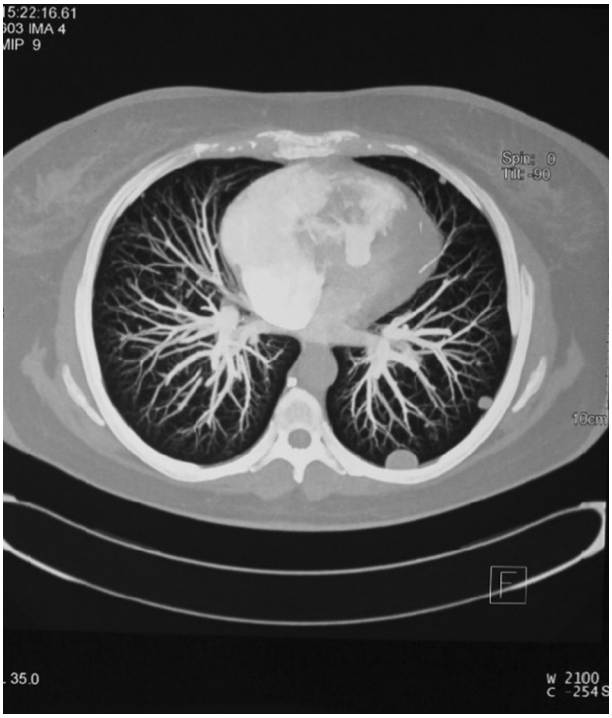


Figure 2. Tumoral Masses in the Right Atrium, Coronary Sinus, Right Ventricle, and Pulmonary Outlet Tract (Computed Tomographic Angiography)

Median sternotomy was performed and cardiopulmonary bypass instituted. Right atriotomy revealed a large mass, attached to the septal leaflet of the tricuspid valve, and a mass in the coronary sinus. The ventricular mass, which originated from the posterior ventricular wall, extended to the chordae tendineae. A large red mass was seen in the right ventricle's outflow tract. The mas was completely resected (Figure 3).

The patient was extubated in shorter than 10 hours of surgery. She recovered without complications and was discharged from hospital nine days later. Histopathologic examination revealed a metastatic ESS, which was confirmed by Immunohistochemistry (IHC). The estrogen receptor (ER), progesterone receptor (PR), and CD10 were positive in the tumor cells. She was referred to the oncologist for adjuvant therapy. After 28 months, follow-up examination revealed no abnormality. All patient information was kept confidential and for publication of the article, written informed consent was obtained.

3. Discussion

The LG-ESS is generally a low-grade malignant neoplasm with an indolent clinical course. Patients with FIGO stage I and II tumors having a five-year survival rate of more than 90%. In contrast, advanced-stage tumors have a five-year survival rate of 40% to 50% (1).

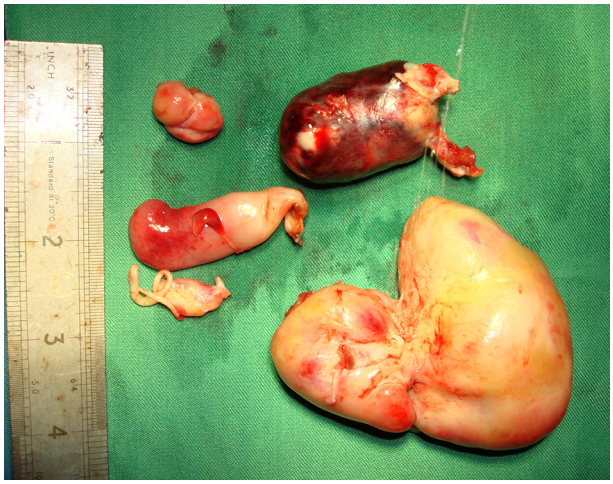
Recurrences are common, occurring in one-quarter to one-half of patients, and the risk is greater in those with more advanced-stage disease (3). Late recurrence and distant metastases may occur. These tumors usually grow slowly and recurrences occur late. Prolonged survival and even therapy are common after surgical resection of the recurrent or metastatic lesions (2).

The current standard of therapy for stage I ESS is hysterectomy and bilateral salpingoophorectomy and tumor debulking is reserved for advanced stages (3). Although chemotherapy has generally shown to be ineffective, success has been showed with adjuvant radiation therapy (1).

The majority of the cases are intrauterine. However, rarely, LG-ESS may present at distant sites such as the ovary and lung (1), and its prognosis is related to extrauterine development (4). Although the ESS tends to spread throughout the lymph nodes and veins, it rarely involves the large vessels. Lymph node metastases occur in up to 30% of cases, and spread to para-aortic nodes has been reported (5-7).

In English literature, only 19 cases of intracavitary extension to the heart from a LG-ESS with successful excision have been reported (8). Our case is the first of its kind with the involvement of the complete right side of the heart and lung metastasis without caval involvement. The pathogenesis of these lesions remains unknown, but exposure to tamoxifen and unopposed estrogens has been implicated in some cases (9). The LG-ESS is more common than ESN, and shares a similar history and clinical presentation with the majority occurring in premenopausal women. A few cases have been reported in younger women or young adolescents (1, 4).

Figure 3. Large Multiple Excited Masses From the Right Ventricle and Right Outflow Tract



Metastatic endometrial stromal sarcoma was confirmed by histopathologic examination and immunohistochemistry.

A useful initial IHC panel is CD10, desmin, ER, and PR. Although CD10 is uniformly expressed in endometrial stroma, it is nonspecific and is identified in a variety of tumors including smooth muscle neoplasms (1).

The clinical presentation of these lesions depends on the localization of the tumor; they tend to present with progressive dyspnea, arrhythmia, chest pain, lower limb edema, and pericardial effusion. Intracardiac metastases are usually discovered during investigations for heart failure or at autopsy (10, 11). Sudden cardiac decompensation or death from the obstruction of the heart valve can occur. The diagnosis is usually confirmed by imaging modalities such as echocardiography and CT. Extensive radical surgery is the best option for treatment because it can potentially improve recurrence-free survival. A panel of IHC stains is used to confirm the diagnosis. The prognosis may be poor, but prolonged survival, up to five to six years, has also been reported (12).

Women with metastatic uterine sarcomas have an overall poor prognosis. Surgical approach to cardiac metastasis offers the possibility of an excellent long-term prognosis in this disease. This isolated metastasis supports a hematogenous mechanism of spread for the ESS.

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Authors' Contributions

Study concept and design: Behnam Shakerian and Mohammad Hossein Mandegar. Acquisition of data: Behnam Shakerian and Bahieh Moradi. Analysis and interpretation of data: Behnam Shakerian and Mohammad Hossein Mandegar. Drafting the manuscript: Bahieh Moradi. Critical revision of the manuscript for important intellectual content: Bahieh Moradi, Behnam Shakerian, and Farideh Roshanali. Administrative, technical, and material support: Behnam Shakerian and Bahieh Moradi. Study supervision: Behnam Shakerian.

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