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

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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
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 mass 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 extraterine 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).
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.

References

1. Conklin CM, Longacre TA. Endometrial stromal tumors: the new WHO classification. Adv Anat Pathol. 2014;21(6):383–93.
2. Kashi Z, Meibodi SJA, Emadi SF. Endometrial stromal sarcoma presented with galactorrhea. Iranian Red Crescent Med J. 2009;11(3):334.
3. Leath C3, Huh WK, Hyde JJ, Cohn DE, Resnick KE, Taylor NP, et al. A multi-institutional review of outcomes of endometrial stromal sarcoma. Gynecol Oncol. 2007;105(3):30–4.
4. Tahmasebi M, Cina M. Bilateral endometrial stromal sarcoma originating from ovarian endometriosis: a case report. Iranian J Radiol. 2009;6(1):33–6.
5. Ma SK, Zhang HT, Wu LX, Liu LY, Li B. Treatment and prognosis of low-grade malignant endometrial stromal sarcoma. Zhonghua Zhong Liu Za Zhi. 2007;29(3):74–8.
6. Montag TW, Manart FD. Endolymphatic stromal myosis: surgical and hormonal therapy for extensive venous recurrence. Gynecol Oncol. 1989;33(2):255–60.
7. Riospel J, Plante M, Renaud MC, Roy M, Tetu B. Lymph node metastases in low-grade endometrial stromal sarcoma. Gynecol Oncol. 2005;96(2):402–6.
8. Renzulli P, Weimann R, Barras JP, Carrel TP, Candinas D. Low-grade endometrial stromal sarcoma with inferior vena cava tumor thrombus and intracardiac extension: radical resection may improve recurrence free survival. Surg Oncol. 2009;18(3):57–64.
9. Beer TW, Buchanan R, Buckley CH. Uterine sarcoma following tamoxifen treatment. J Clin Pathol. 1995;48(6):596.
10. Harting MT, Messner GN, Gregoric ID, Frazier OH. Sarcoma metastatic to the right ventricle: surgical intervention followed by prolonged survival. Tex Heart Inst J. 2004;31(3):53–5.
11. Fernando Val-Bernal J, Hernandez-Nieto E. Symptomatic intracavitary (noninvasive) cardiac metastasis from low grade endometrial stromal sarcoma of the uterus. Pathol Res Pract. 1999;195(10):717–22.
12. Yokoyama Y, Ono Y, Sakamoto T, Fukuda I, Mizunuma H. Asymptomatic intracardiac metastasis from a low-grade endometrial stromal sarcoma with successful surgical resection. Gynecol Oncol. 2004;92(3):999–1001.