Resection of an Endocardial Uterine Sarcoma Metastasis to the Tricuspid Valve Apparatus with Valve Repair

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Central Message:
We present the first reported case of a successful resection of a tricuspid valve metastatic endometrial stromal sarcoma lesion with valve preservation and repair.

Abstract

Isolated metastasis of a uterine stromal sarcoma to the cardiac valves is extremely uncommon. We describe a case of successful resection of a tricuspid valve metastatic lesion with valve preservation and repair. We have not encountered a similar case in the literature.
Introduction
Metastatic tumors of the heart are not uncommon in patients with known malignancies. Endocardial location of metastatic disease is much less common compared to the epicardium and pericardium (1, 2). The most common metastatic tumors are melanoma, breast, lung, and gastrointestinal malignancies. Metastatic uterine malignancy of the heart is exceptionally rare. We describe a case of a successfully resected solitary metastasis from a uterine endometrial stromal sarcoma (ESS) to the tricuspid valve anterior leaflet subvalvular apparatus. Resection of the tumor required excision of part of the papillary muscle and chord, with neochordal reconstruction of the anterior leaflet. To our knowledge, this is the first report of such a case. The Institutional Review Board (IRB) of the University of Iowa reviewed the project and considered this case report to be exempt from the application process. The patient provided written consent for publication.

Case Description
A 69-year-old female underwent a total laparoscopic hysterectomy and bilateral salpingo oophorectomy for uterine sarcoma. Pathology was reported as a 9.8 cm, low-grade estrogen (ER) and progesterone (PR) receptors positive endometrial stromal sarcoma with necrosis, lymphovascular invasion, and negative resection margins. One year later, the patient underwent an upper vaginectomy for a focus of microscopic recurrent sarcoma, which was resected completely. The patient did not receive adjuvant chemo or immunotherapy with plans for close clinical follow-up.

During follow-up at 8 months, an intracardiac mass was noted on chest computed tomography (CT). Cardiac CT demonstrated a large heterogeneous mass in the right ventricular outflow tract (RVOT). At least two large lobes were seen that measured 1.2 × 1.2 cm and 1.7 × 0.9 cm. The density of the mass suggested that it was a tumor rather than a thrombus (Fig. 1). A transthoracic echocardiogram confirmed the presence of the mass and revealed mild to moderate tricuspid regurgitation. Bilateral ventricular systolic function was preserved. Additional workup did not reveal any evidence of metastatic disease. The patient did not have any arrhythmias or heart failure symptoms.

The patient underwent excision of the intracardiac mass via median sternotomy using cardiopulmonary bypass (CPB). Because of the complexity of the repair and the need for good oncologic resection, we decided to do the procedure under cardioplegic arrest, this was done in an antegrade fashion. The tumor was found to involve the subvalvular apparatus, including papillary muscle and chords to the anterior leaflet of the tricuspid valve (Fig. 2A). Excision of part of the papillary muscle and chords was performed via right atriotomy. After the tumor was resected with the attached chordal structures, a 4-0 Gore-Tex (W. L. Gore & Associates, Inc, USA) stitch was attached in a figure of eight fashion to an adjacent papillary muscle to support the leaflet edge of the anterior leaflet close to the antero-septal commissure. The neo-chordal length was adjusted to the height of the adjacent septal leaflet chords and tied. A commissuroplasty (antero-septal) was performed with a 5-0 polypropylene suture. Commisuroplasty was needed because of the proximity of the resected chords to the commissure with no additional chordae present to support it. This would prevent commissural regurgitation and help stabilize the close by artificial chords. The valve was tested with saline and was competent except for a central leak. A 28 Tri-Ad ring was utilized for stabilization of the annulus (Fig. 2B). An intraoperative transesophageal echocardiogram demonstrated resolution of the tricuspid regurgitation after repair (Sup.
video 1). CPB time was 125 min and cross clamp time was 88 min.

Pathologic examination of the excised mass was consistent with metastatic endometrial stromal sarcoma (Sup. Figs. 1A and 1B). The postoperative course was uneventful, and the patient was discharged on postoperative day 3. Given the positive ER/PR status of the tumor, the patient was managed with hormonal therapy (Letrozole) after resection of the metastasis. At her 6- and 12-month follow-up visits, the patient was doing well. Imaging demonstrated no recurrent disease. Echocardiogram surveillance showed no recurrent intracardiac mass and no tricuspid valve regurgitation. Postoperatively, the patient remained in normal sinus rhythm.

Comment

While primary cardiac tumors are rare (0.01–0.1% on postmortem analysis), metastases to the heart in patients with known malignancies are not uncommon (1, 2). The most common metastatic tumors are melanoma, breast, lung, and gastrointestinal malignancies. The endocardial location of metastatic disease in the endocardium is much less common compared to in the epicardium or pericardium (1).

An ESS is a rare tumor of mesenchymal origin estimated to comprise 1% of all uterine malignancies and approximately 10% of uterine sarcomas (3). These tumors are classified as endometrial stromal nodules, low-grade, high-grade, and undifferentiated tumors. ESS are indolent hormonally sensitive tumors with a 5-year survival rate of 66-90% depending on the grade. Undifferentiated ESSs have aggressive biologic behavior and an estimated 5-year survival of 25-55% (4). Recurrent disease is seen most commonly in the pelvis and abdomen. Distant metastasis usually occurs in the lungs. The standard treatment for ESS is total hysterectomy with bilateral salpingo-oophorectomy. The therapeutic value of lymphadenectomy in the management of stromal sarcomas is not clear but may have prognostic significance.

Metastatic ESS to the heart is exceptionally rare. There are several reports of ESSs presenting as an intracaval mass extending into the right atrium mimicking thrombus, with only four reported resections (5). These intracardiac extensions may not be adherent to the cardiac wall and can be successfully managed with a staged excision using transcaval extraction and a cardiotomy. Hematogenous metastasis may result in isolated intracardiac lesions. Only a handful of reports describing the resection of metastases located in the right atrium, right ventricle, or involving the valvular apparatus could be found (S1). Available reports have not described a negative resection margin. We found only one report describing De Vega suture anuloplasty of the tricuspid valve with a 3-month follow-up (S2). Similarly, only scant surgical experience with the management of intracardiac metastatic uterine leiomyosarcoma has been described in the literature (S3).

To our knowledge, the case of our patient is the first described case of papillary muscle and chordal resection and neochordal reconstruction after resection of an isolated right heart ESS metastasis with a negative margin. Our case and its one-year-long follow-up with a successful oncologic outcome and functional results confirms that isolated metastatic disease to the right heart can be successfully managed and result in a good outcome. We recommend that patients with an isolated metastatic uterine ESS disease involving the valvular apparatus be evaluated for possible surgical resection and reconstruction.
Figure 1. Cardiac computed angiography demonstrates a 21 mm × 19 mm multilobar mass attached to the right ventricular side of the anterior interventricular septum protruding into the RVOT just below the pulmonary valve.

Figure 2.
(A) Intraoperative photograph through the right atriotomy demonstrating bilobar mass attached to the subvalvular apparatus of the anterior leaflet.
(B) Intraoperative photograph demonstrating completed tricuspid annuloplasty repair.

Supplemental Figure 1.
(A) Microscopy of primary low-grade endometrial stromal sarcoma with lymphovascular invasion (hematoxylin-eosin, magnification × 40).
(B) Microscopy of the tricuspid valve involved by metastatic endometrial stromal sarcoma (hematoxylin-eosin, magnification × 25).

Video 1. Preoperative echocardiogram demonstrates the mass and mild to moderate tricuspid regurgitation. Post resection intraoperative echocardiogram demonstrates resolution of the tricuspid regurgitation.

References

Supplemental References