Case Reports

Endometrial Stromal Sarcoma

Metastatic from the Uterus to the Inferior Vena Cava and Right Atrium

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Endometrial stromal sarcoma metastases usually occur within the pelvis and rarely involve the great vessels or the heart. We present the case of a 55-year-old woman who was referred for endovascular therapy to treat presumed thrombosis of the inferior vena cava. The suspected thrombus was recalcitrant to endovascular removal with use of an AngioVac venous drainage device. Results of an intraprocedural transvenous biopsy revealed the mass to be the intravascular extension of an endometrial stromal sarcoma. The patient underwent surgical excision of the tumor, and, shortly thereafter, a hysterectomy and salpingo-oophorectomy. This complex case highlights both the rarity of malignancy masquerading as caval thrombus and the importance of multispecialty collaboration. ***(Tex Heart Inst J 2015;42(6):558-60)***

Key words: *Heart atria/surgery; heart neoplasms/secondary/surgery; neoplasm invasiveness; sarcoma, endometrial stromal/complications/diagnosis/pathology/surgery; treatment outcome; uterine neoplasms/pathology; vascular access devices; vena cava, inferior/pathology/surgery*

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The malignancy that most often extends into the inferior vena cava (IVC) is renal cell carcinoma.[1] We describe the case of a patient who presented with a presumed IVC thrombus that extended into the right atrium. After the failure of endovascular removal attempts with use of an AngioVac® venous drainage device (AngioDynamics; Latham, NY), a transvenous biopsy specimen yielded a low-grade endometrial stromal sarcoma. We also discuss the use of the AngioVac and the multidisciplinary collaboration involved in our patient's treatment.

Case Report

In August 2013, a 55-year-old woman was transferred to our hospital with symptomatic pulmonary embolism (PE). Her medical history included hypothyroidism and postmenopausal spotting. Although the patient was normotensive, she needed 2 L/min of oxygen through a nasal cannula. She was orthopneic and had difficulty walking. The symptoms of PE persisted despite standard systemic anticoagulation with infused intravenous heparin. A computed tomographic (CT) pulmonary angiogram from the referring institution revealed a filling defect—initially thought to be thrombus—in the IVC, extending into the right atrium (Fig. 1). A transthoracic echocardiogram confirmed these findings (Fig. 2). Additional CT of the abdomen and pelvis showed that the suspected thrombus originated in the right common iliac vein. No gross visceral abnormality was seen; the only notable finding in the pelvis was an enlarged, fibroid uterus.

On hospital day 3, in an effort to extricate the suspected thrombus in a minimally invasive manner, the cardiothoracic and interventional radiology teams undertook endovascular thrombectomy with use of the AngioVac. From a right internal jugular approach, the right atrial mass was engaged by the AngioVac cannula several times under fluoroscopic and transesophageal echocardiographic observation; however, the mass could not be retrieved through the cannula. The operators, thinking that the mass could be cleared from another approach, placed the AngioVac cannula into the IVC through the left common femoral vein, but this too was unsuccessful. Given the adherent nature of the suspected thrombus, a transvenous biopsy was performed. Pathologic analysis of the biopsy specimen revealed a tumor embolus.

On hospital day 5, the cardiothoracic and vascular services teams performed open surgical excision of the mass. After a median sternotomy, cardiopulmonary bypass was started, the patient was systemically cooled to 16 °C, and heparin was administered. First, the right atrium was opened. The tumor mass was firmly attached to the mass

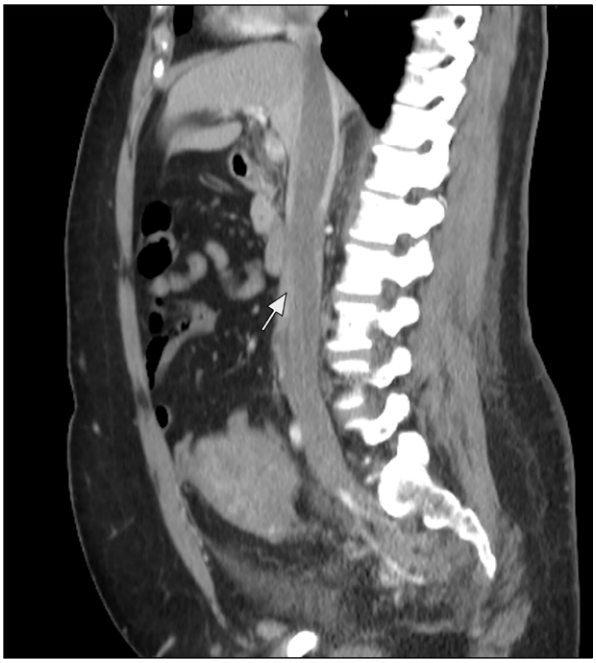

Fig. 1 *Computed tomogram (sagittal view) of the chest, abdomen, and pelvis shows a mass within the inferior vena cava, extending into the right atrium.*

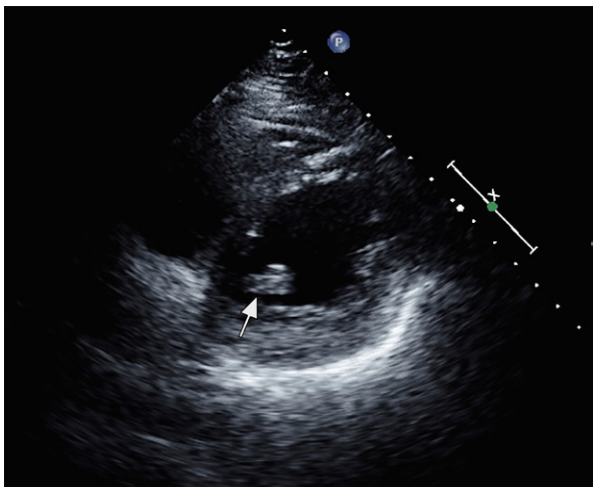

Fig. 2 *Transthoracic echocardiogram shows a mobile thrombus (arrow) within the right atrium.*

within the IVC and could not be easily extricated. The IVC was opened at the level of the renal veins, and the tumor was removed with careful sharp and blunt dissection. (Of note, the renal veins were free of tumor.) The IVC was then opened inferiorly toward the bifurcation into the common iliac veins, and more tumor was removed. Next, attention was turned to the right main pulmonary artery. Tumor mass was identified and removed from the trifurcation branches. The mass that extended from the right atrium to the right common

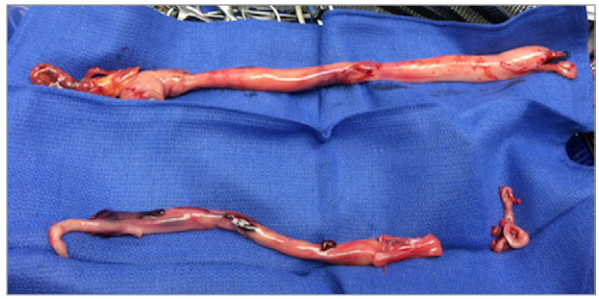

Fig. 3 *Photograph shows gross specimens of the resected tumor thrombus.*

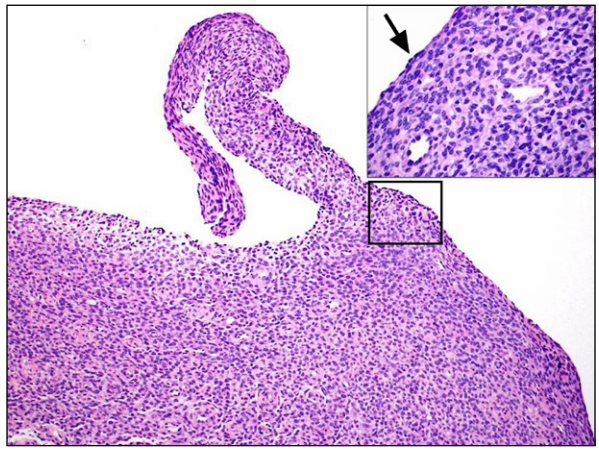

Fig. 4 *Photomicrograph shows a uniform population of small cells with eosinophilic cytoplasm but without associated blood clot (H & E, orig. ×10). At higher power, round-to-oval nuclei with fine chromatin and surface endothelialization are seen (arrow) (H & E, orig. ×40).*

iliac vein was successfully removed en bloc (Fig. 3). The total time of circulatory arrest was 41 min.

The largest tumor fragment (from the IVC) was 33.5 cm long. On cut section, the specimen consisted of tan-red variegated, firm tissue without evidence of superimposed thrombus. Histologic sections showed a population of small cells with moderate eosinophilic cytoplasm, round-to-oval nuclei, fine chromatin, and associated delicate vessels (Fig. 4). Mitosis was focally increased (<10 figures/high-power field), and areas of necrosis were easily identified. Immunohistochemical stain revealed that the cells were reactive to cytokeratin AE1/AE3, estrogen receptor, progesterone receptor, and CD10; and nonreactive to desmin, *PAX8*, HMB-45, melan-A, and inhibin. The overall immunohistochemical and morphologic profile was that of low-grade endometrial stromal sarcoma.

On hospital day 7, the gynecology service performed a total abdominal hysterectomy and bilateral salpingo-oophorectomy. After an 81-day hospitalization (extended partially because of rehabilitation), the patient was discharged with instructions to take 1 mg/d of sys-

temic anastrozole (an aromatase inhibitor that prevents the conversion of androgen to estrogen). Six months after surgery, the patient sustained hydronephrosis of the right kidney, related to a benign stricture within her mid right ureter. The urology service placed ureteral stents. One year postoperatively, the patient had no clinical or radiologic sign of tumor recurrence, had recovered fully from her surgeries, and had resumed activities of daily living.

Discussion

The AngioVac has been effective in clearing femoral, iliac, and IVC thrombus.[2] The device consists of a large-bore cannula with an aspiration cannula that is attached to an extracorporeal bypass circuit and a reinfusion cannula. The U.S. Food and Drug Administration has approved the AngioVac to "remove undesirable material during the performance of extracorporeal bypass." Its use to retrieve caval thrombus, intracardiac thrombus and masses, and pulmonary emboli is well described.[2-4] The AngioVac has been used to prevent PE from tumor thrombus[5] and can be used in patients who have contraindications to thrombolytic therapy or who are at high risk for surgical thrombectomy.

Our patient was initially thought to be at moderate-to-high risk for open surgical thrombectomy. However, because of the adherent nature of this IVC tumor, use of the AngioVac was unsuccessful. After collaboration between our cardiothoracic surgery and vascular surgery teams, the patient was taken for open surgical resection of the tumor. When the origin of the mass was identified, hysterectomy and bilateral salpingo-oophorectomy were performed.

Endometrial stromal sarcoma constitutes approximately 0.2% to 1% of all uterine malignancies and approximately 10% of uterine sarcomas; its annual incidence is approximately 2 in 1,000,000 women.[6,7] Its clinical recognition can be difficult, and it is often mistaken for a leiomyoma until a true diagnosis is made postoperatively. Histologic examiners miss the diagnosis of early-stage, low-grade endometrial stromal sarcoma at a reported rate of up to 40%, which results in therapeutic delay.[8]

Endometrial stromal sarcoma is classified as low-grade or high-grade, on the basis of mitotic activity. Low-grade tumors have fewer than 10 mitotic figures per high-power field,[9] a 5-year survival rate of 80% to 100%, and approximately a 50% rate of recurrence, typically after a long latency period. In one large series, the interval before recurrence varied from 3 months to 23 years, with a median interval of 3 years.[4] Patients with high-grade endometrial stromal sarcoma have a 5-year survival rate of 25% to 55%.[6]

The most frequently reported sites of metastases of the low-grade subtype are the vagina, pelvis, and peritoneal cavity. Endometrial stromal sarcoma tends to spread throughout the lymph nodes and venous system but rarely involves the large vessels or the heart.[9] In general, intracardiac extension of tumors is rare. Most cases derive from renal cell carcinoma, nephroblastoma, colon adenocarcinoma, melanoma, hepatocellular carcinoma, or bronchogenic carcinoma.[9] Only 19 cases have been reported of advanced low-grade endometrial stromal sarcoma that has invaded the great vessels with formation of an IVC tumor.[6]

This case highlights the importance of a multidisciplinary approach in treating this rare cardiovascular pathologic condition.

Acknowledgments

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