Uterine Smooth Muscle Tumor of Uncertain Malignant Potential (STUMP) Extending via the Inferior Vena Cava into the Right Atrium

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Abstract

We present a 29-year-old female, who was hospitalized because of dyspnea and generalized edema. The patient had a history of splenectomy due to resistant idiopathic thrombocytopenia purpura (ITP) and also a history of smooth muscle tumor of the uterus with uncertain malignant potential (STUMP). Echocardiography revealed large pericardial effusion and an elongated mass inside the inferior vena cava and right atrial cavity. The patient underwent only cardiac surgery procedure under monitoring by transesophageal echocardiography. The inferior vena cava and right atrium were exposed and the large mass was removed. Histological examination revealed a spindle cell tumor. ***(Iranian Heart Journal 2012; 13(2):54-58).***

Keywords: Cardiac tumor■ Uterus leiomyoma■ Transesophageal echocardiography■ Inferior vena cava

Introduction

Patients with tumors sometimes develop complications in the cardiovascular system (1). These complications can occur as a result of a locally invasive disease or distant spread (1). Primary tumors of the heart are uncommon and are usually benign (1).

In contrast, direct extension of tumors, hematogenous spread, and retrograde lymphatic extension to the heart are common (1). Tumors may prorogate within blood vessels returning to the heart, most commonly originating from abdominal tumors via the inferior vena cava to the right-sided heart (2).

Renal cell carcinoma, adrenal carcinoma, hepatocellular carcinoma, and uterine leiomyosarcoma are the most common tumors that reach the heart in this manner (2). In most cases, the initial diagnosis of these tumors is made after the detection of a right atrial mass on echocardiography (3). The tumors that involve the right side of the heart may require transesophageal echocardiography (TEE) for diagnosis (3). Diagnostically, TEE is useful because it provides excellent visualization of venous inflow to the heart (2). In cases of intraluminal leiomyosarcoma, multiple points of attachment in the inferior vena cava may help distinguish this tumor from renal cell carcinoma or hepatoma (4).

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Patients with tumors may present acutely with cardiac symptoms or catastrophic hemodynamic instability, but presentation is more commonly nonspecific (5).
Initial diagnosis of a cardiac tumor is best made with echocardiography, but a more complete assessment of the extent of local and regional disease using CT and MRI is required for curative management (5).

Case Presentation

We describe a 29-year-old woman with a history of splenectomy four months previously due to resistant idiopathic thrombocytopenic purpura (ITP) and also a history of a smooth muscle tumor with uncertain malignant potential (STUMP) of the uterus. Excisional biopsy of the uterine mass had been done for her about 10 months previously.
She was hospitalized in our center with dyspnea on exertion (functional class III) and generalized edema of50 days' duration without acute exacerbation.
On admission, the patient's general appearance showed facial puffiness, neck engorgement, generalized edema, and abdominal protrusion due to large ascites. Her vital signs revealed a blood pressure of 70mmHg on pulse, a pulse rate of 120 beats per minute, and a respiratory rate of 16 per minute.
The electrocardiogram showed low voltage QRS and non-specific ST-T changes.
Laboratory findings revealed the following: ESR=9; WBC=15000; PMN=95%; Plat=145000; Hb=7.6 g/dl; normal creatinine, urea, sodium, potassium, and sugar; mildly increased AST; and increased total bilirubin (3.7 mg/dl) with normal direct bilirubin (0.8 mg/dl).
Echocardiography revealed massive pericardial effusion without tamponade physiology, and a large elongated mass inside the inferior vena cava and right atrium, passing into the tricuspid orifice, causing severe functional tricuspid stenosis(mean gradient=13 mm Hg), and also entering the right ventricular outflow (Figures 1a,1b).
CT scan demonstrated ascites in the abdominopelvic cavity and a cauliflower mass at the right side of the uterus, which suggested subserosal myoma. A mass in the right atrium and a mass lesion producing an intraluminal filling defect with adherence to the wall in the infra-hepatic and intra-hepatic parts of the inferior vena cava were noted, suggesting a tumoral lesion such as leiomyoma or leiomyosarcoma.
The patient underwent surgical operation about 5 hours after admission and a large mass (10 × 8× 6cm) that extended firmly into the inferior vena cava was found and excised, including that from the vena cava. The cardiac operation was monitored via TEE.
Gross pathology revealed a solid and creamy mass with small cyst-like spaces or foci of hemorrhage (Figure 2).
Microscopic examination showed a neoformative tissue, composed of relatively monomorphic cells with plump spindle-shaped or oval nuclei, rare mitotic or bizarre cells, and foci of tumoral necrosis, suggestive of a spindle cell tumor (Figure 3).

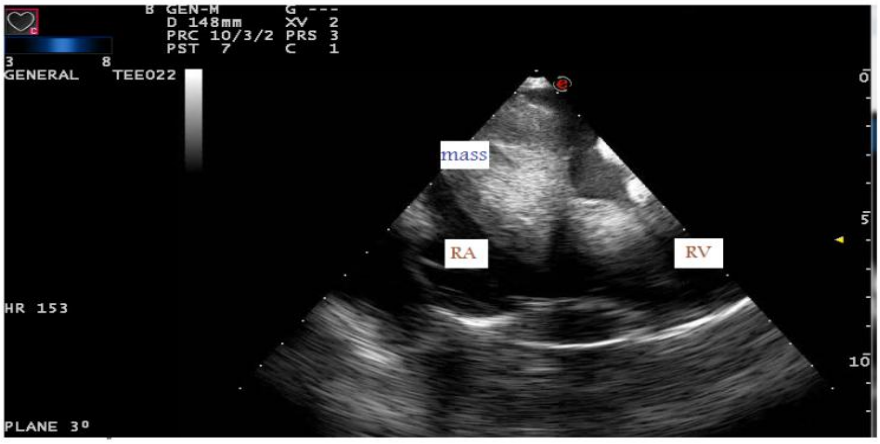

Figure 1a: Two-dimensional transesophageal echocardiography in plane about 0° shows a huge mass in the right atrium, which protrudes into the right ventricle from the tricuspid valve. RA: right atrium. RV: right ventricle

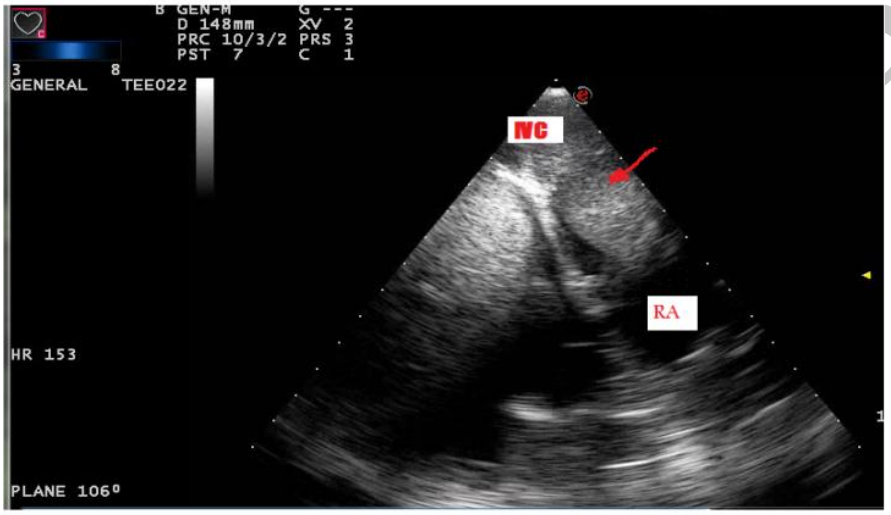

Figure 1b: Two-dimensional transesophageal echocardiography in plane 100° shows a huge mass (red arrow), which protrudes into the right atrium through the inferior vena cava. IVC: Inferior vena cava, RA: right atrium

Figure 2: Gross pathology shows a solid and creamy mass with small foci of hemorrhage

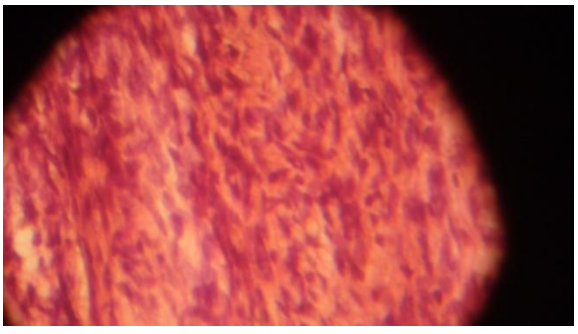

Figure3: Microscopic examination shows relatively monomorphic cells with plump spindle-shaped or oval nuclei, rare mitotic or bizarre cells, and foci of tumoral necrosis, suggestive of a spindle cell tumor.

Discussion

The World Health Organization (WHO) has classified uterine tumors that cannot be histologically diagnosed as unequivocally benign or malignant and as smooth muscle tumors of uncertain malignant potential (STUMP). The WHO's classification of mesenchymal tumors describes STUMP as an intermediate tumor between a benign leiomyoma and a malignant leiomyosarcoma. Compared to malignant leiomyosarcoma, STUMP has a superior prognosis, but the biological potential of the tumor remains unclear. Lymphogenic and hematogenic dissemination seems possible even after a long period of time (6).

Previously, it was believed that leiomyosarcoma was a high-grade sarcoma associated with poor prognosis even if at stage I. Conversely, most tumors classified as STUMP have been associated with favorable prognosis. (7). We herein presented a case with STUMP, which progressed to poor prognosis.

Our case illustrates that rare smooth muscle tumors of the uterus may have metastatic potential without obvious malignant morphologic features and, therefore, require a close follow-up.

In some reported cases, both median sternotomy (cardiotomy) and laparotomy have been drawn upon to withdraw one portion of the mass from the right atrium and another from the abdominal inferior vena cava (8).

With respect to the emergent condition of our patient, we chose only sternotomy to prevent pulmonary embolization of the right atrium mass or sudden death because of incursion into the arteriovenous orifice. We intend to use periodic postoperative ultrasonic or MRI to detect the growth of the residual intravenous tumor.

In another case presented by Okamura H et al., the tumor partially was removed from the right atrium and the inferior vena cava via the right atrium using cardiopulmonary bypass. The residual tumor was not found after 25 months of follow-up (9).

We found no correlation between idiopathic thrombocytopenia purpura (ITP) and the presence of the heart tumor and we think this coincidence is an incident.

Conclusion

We here in presented the first description of a case of uterine STUMP, which extended to the heart via the inferior vena cava and with a recurrence form of a spindle cell tumor.

This case highlights the importance of echocardiography in revealing the cardiac involvement by the tumor via the inferior vena cava and in planning the operative procedure.

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