A Road to the Heart From Uterine Closet: A Case Report

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Abstract- Angiosarcomas of the uterine is a malignant and poor prognostic tumor and can either be a primary sarcoma or arising secondary to radiotherapy of a more complex tumor. Primary uterine angiosarcomas are exceptional and probably arise from embryonic vascular remnants, teratoma or from the rich uterine vasculature. We reported a rare case of primary angiosarcoma of uterine that at the time of diagnosis presented with sign and symptom of local and distant metastasis. The patient presented with dyspnea, chest pain, and history of vaginal bleeding and pelvic pain. The physical exam revealed pallor, prominent jugular pulse pressure, a palpable fixed mass in the pelvic however vaginal exam was unremarkable. Transthoracic echocardiography (TTE) revealed massive pericardial effusion and also a large mass in the right atrium. The abdominal ultrasound showed echogenic and poor echogenic segments in uterine mass combined with central necrosis. The patient underwent total hysterectomy and Bilateral salpingo-oophorectomy followed by radiotherapy and adjuvant chemotherapy. The patient underwent open heart surgery with resection of cardiac mass and further received a four cycle of radiotherapy (50 MG) to the mediastinum. The further follow-up (6 month) revealed no recurrence of tumor in a mediastinum. However, patient died from metastasis to the liver and its hepatic failure sequels.

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Keywords: Uterine sarcoma; Metastasis; Cardiac involvement

Introduction

Uterine sarcomas as a soft tissue sarcoma are uncommon malignant tumors that composed 1% of genital organ malignancies and approximately 5% of uterus cancers (1). Uterine angiosarcoma is a tumor in which Malignant cells arise in the endothelial lining of uterus blood vessels or other supporting tissue of uterus such as ligament or adenex. In opposed to others common uterine sarcomas such as leiomyosarcoma, alveolar sarcoma, liposarcoma, spindle cell sarcomas and Ewing sarcoma angiosarcoma of uterine is exceedingly rare. However aforementioned sarcomas had risk factors such as the history of radiotherapy to the pelvis or tamoxifen therapy for breast cancer, but angiosarcoma has not the known risk factor for its development (2). Angiosarcoma accounts for less than 1 percent of all gynecological malignancy. Uterine angiosarcomas are exceptional and have a poor prognosis. however, uterine sarcomas are the most common sarcoma of genital organs, but angiosarcomas of the uterine comprise less than 0.5 percent of uterine malignancies, mostly raised from others malignancy like epithelial carcinoma or carcinosarcomas (3). Angiosarcomas comprise only 2 percent of all soft tissue sarcomas. These tumors may originate in the coetaneous vascular channel, muscles, soft tissue, the breast and the skin angiosarcoma most commonly arises from the skin of the head and neck region in elderly. Delay in diagnosis, deep tissue invasion, and early metastasis caused angiosarcoma of deep soft tissue as uterine, a poor prognosis malignant tumor. Gynecology origin of angiosarcoma have also been reported in the the vagina, vulva, cervix ,ovary and rarely uterine, and careful literature searching revealed only fewer than 12 reported cases in the Medline.

Case Report

A 48-year-old woman, para 3+0, referred to our center with dyspnea, chest pain, malaise, abdominal pain and with a TTE that showed a large mass in the right atrium and massive pericardial effusion. The physical exam revealed mucous membrane pallor, respiratory

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distress, and fingertips cyanosis. Chest auscultation showed reduced lower left chest respiratory sounds. Heart sound was reduced, and jugular vein was prominent. The patient's consciences were normal and no any neurologic deficit was found, in the abdominal exam, a fixed mass with irregular counter was found in the region, and moderate ascetics were also detected in the peritoneal cavity. She gave a history of vaginal spotting for the last two months. Her menstrual cycles were unremarkable. The blood exam showed hemoglobin of 8 mg/dl. Liver function test was in upper normal levels, and renal dysfunction was detected by abnormal limits of creatinine (CR=2.5 mg/dl). TTE showed severs pericardial effusion and a large mass was detected in the right atrium (Figure 1). An ultrasound evaluation of abdominal organs revealed also a large-sized uterus displaced to the left side by a unilateral mass, with maximum dimension of 7 cm. Large free fluid was seen in the peritoneal cavity. Abdominal ultrasound examination of the liver and the retroperitoneal organ was normal. The primary clinical diagnosis was an invasive uterine tumor with distant metastasis to the right atrium. With regard to priority of surgery and symptom of severe pericardial effusion and possibility of right atrial mass emboli to the right ventricle and pulmonary artery, the patient scheduled for cardiac surgery. The patient underwent midline sternotomy with aortic and bicaval cannulation. The ascending aorta cross-clamped and with antegrade cardioplegia infusion, and cardiac arrest, right atrium was opened and a large infiltrative mass attached to the right atrial external wall resected with a small part of the atrial wall (Figure 2,3). This defect repaired with the fresh pericardial patch. The pericardial effusion was bloody and developed by tumor invasion to atrial wall and pericardium. The postoperative course of cardiac surgery was uneventful, and the patient moved to ward to continue evaluation of pelvic mass and ascetic fluid and further surgery. The ascetic fluids were sent for searching of malignant cells with the negative finding.Two weeks later the patient prepared for laparotomy. Frozen sectioning of uterine mass, exhibits it as a necrotic and hemorrhagic legion with negative malignant cells. Based on the pathological finding, benign or malignant nature of mass could not be ascertained by this method. The patient underwent radical hysterectomy with bilateral salpingo-oophorectomy. These specimens sent for pathological evaluation that composed of aforementioned organs. Grossly, the midline uterine mass measured 12 cm×14 cm extended to adenex and had necrotic and hemorrhagic parts, with some solid and cystic areas (Figure 4). Pathological exam of the uterine mass revealed a vascular neoplasm with primitive channels covered with the malignant epithelial cell. Active vasoformation with collateral arbor zing channels and different sizes and contour covered by atypical endothelial cells also found in the microscopic exam. The capsule of the tumor was disrupted and invaded to parauterine organs. The high mitotic count and areas of tumor necrosis were also seen along with the chaotic formation of fused anastomosing vascular parts (Figure 5,6). No primitive cells or teratomatous parts were detected. A facility for CD31, CD34 antigens, Glycogen and mucin staining, were not available in our center. Our patient was diagnosed as having a high-grade angiosarcoma with distant involvement of heart chamber. Our patient was referred to oncology center for further management, and subsequently died with diffuse lung metastasis.

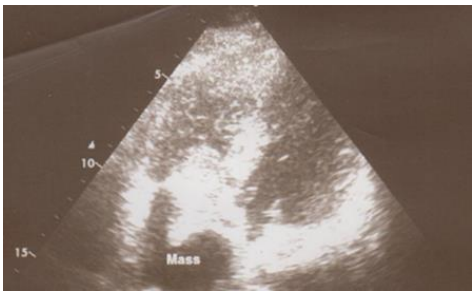

Figure 1. TTE shows a large mass attached to right atrial wall

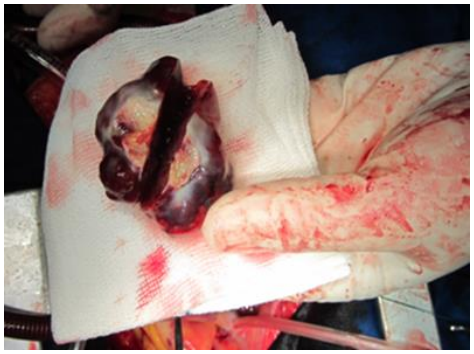

Figure 2. Shows gross view of resected metastatic cardiac tumor

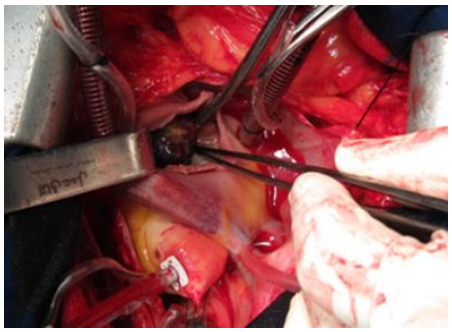

Figure 3. Shows intraoperative view of metastatic uterine angiosarcoma to right atrium

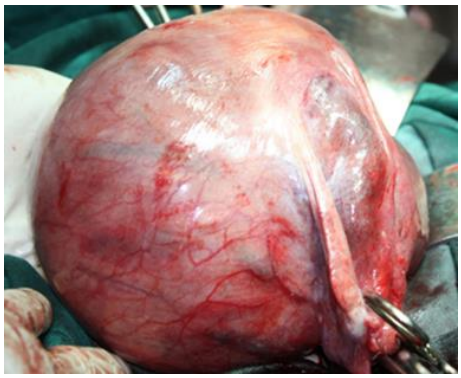

Figure 4. Shows gross pathology of resected angiosarcoma

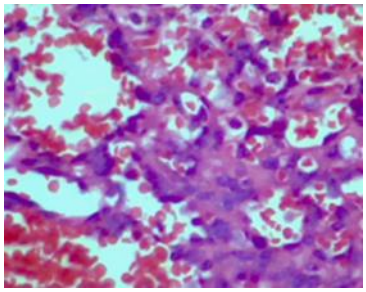

Figure 5. Well-formed vasoformative channels. Hematoxylin and eosin stain, 20×

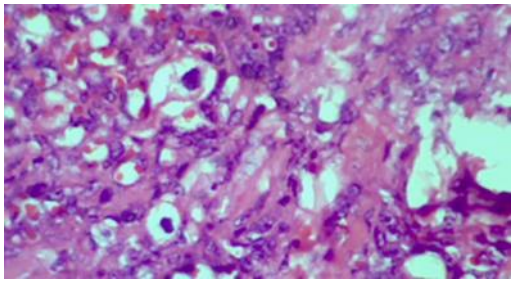

Figure 6. Higher magnification is showing solid areas with nuclear pleomorphism and scattered mitotic figures. Hematoxylin and eosin stain, 40×

Discussion

Uterine angiosarcomas are rare and primary arise from the embryonic vascular remnant of the uterine wall or germ cell tumor with rich vascular supply (4,5-8). The development of malignancy as primary angiosarcoma from a vascular network of uterine is not well-known and its pathogenesis remains unclear. The sarcomatous components of the angiomatous lesion in uterine may reveal itself in the primary uterine tumor or appeared as recurrence of tumor or appeared with diffuse metastases. Malignant changing of some mesenchymal components in teratomas, originated from primary germ cells may transform some primitive cells in yolk sac masses to sarcoma-like angiosarcoma, To date, only less than handful cases of uterine angiosarcoma have been reported in medical literature. In opposing to ovarian angiosarcomas that approximately 30% of them are linked with another malignancy, few cases of uterine angiosarcomas developing from teratomas of the uterine origin have been reported so far. Of these cases of primary uterine angiosarcoma reported in medical literature, only one case originated in a uterine wall mesenchymal cell. One another case of uterine Angiosarcoma was related to the associated epithelial tumor: one case originated from mucinous cystadenocarcinoma and another one from a serous, mucinous tumor of low-grade malignant potential. Uterine Angiosarcomas may be presented as a unilateral, bilateral or diffuse pelvic mass or by sign and symptom of distal metastasis (9,10,11). The tumor can appear at a minimum age of year to a maximum age of 80 years, however, the occurrence of tumor in premenopausal age is exceptional, and no cases have been reported in patients in premenopausal age. However the most common symptom of mass is pelvic pain, but some cases presenting with distant metastases sequels such as hemoptysis, jaundice, neurologic sequels or soft tissue masses. Other clinical features of the tumor include diffuse peritoneal involvement, retroperitoneal invasion, and disseminated intravascular coagulation. Some patients are presenting with ascites or pleural effusion. Angiosarcoma can spread directly by peritoneal invasion, lymphatic spreading, or blood-borne dissemination. Although peritoneal invasion is the most common modality for angiosarcoma dissemination, high tendency of this type of sarcoma by hematogenous metastases caused simultaneous diagnosis of distant dissemination of tumor with primary of the tumor as a common phenomenon. The rich pelvic vein network that related to the great veins caused malignant cells easily reaching to neighboring organs such as liver, kidney or abdominal organs, however, it is unknown how the malignant cell could pursue and bypass the liver filter and reach the cardiac chambers. It seems that malignant cells may bypass the liver by collateral veins and seeds in the heart chambers. In other hand ,malignant cell reaches to lymphatic chains and further to the subclavian veins and the right atrium. Most cases of uterine Angiosarcoma have distant metastasis at the time of diagnosis and rare cases were detected at the early stage as our case that its rapid progression from cardiac surgery to death was less than eight months. Diagnosis of uterine Angiosarcoma is associated with advanced progression of the tumor to a stage of disease that its behavior is highly aggressive and exhibits a poor

response to surgical excision, chemo and radiotherapy ablation (12-14).

Patients with uterine angiosarcomas may concomitantly present with sign and symptom of primary tumor such as pelvic pain, and vaginal bleeding combined with effects of distant metastasis in affected destination organ. However in some patient presenting symptoms related to distant metastases, usually in the lungs. Spreading of malignant tumor beyond the uterine is present at the time of laparotomy in most of the literature cases, with rapid tumor extension within less than a ten months after diagnosis (15,16). Our case with aggressive behavior and advanced stage exhibits poor response to surgery and adjuvant chemotherapy and radiotherapy, with an overall poor prognosis. This tumor has a tendency for combined local invasion and distant metastases, and prognosis is hence poor.

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