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CASE REPORT

Removal of an endometrioid stromal sarcoma from the inferior vena cava and right atrium

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Abstract

Entometrioid stromal sarcomas are seen in extra-uterine as well as extra-gonadal sites and have a strong association with endometriosis. Although having better prognosis than other sarcomas, yet these tumors may relapse (whether local or distant) in up to 56% of cases, even as late as 20 years after surgery. We report a case of a 30-year-old female patient with a mass in the inferior vena cava and right atrium which was surgically removed using cardiopulmonary bypass and deep hypothermic circulatory arrest and turned to be an entometrioid stromal sarcoma. The patient gave a history of endometriosis followed by the appearance of a low-grade ovarian endometrioid stromal sarcoma 4 years before the development of the mass in the IVC and right atrium.

Keywords Entometrioid stromal sarcoma · Cardiac masses · Total circulatory arrest

Introduction

Some abdominal malignancies like renal cell carcinoma and uterine malignancies have been reported to extend or reappear in the inferior vena cava (IVC) and right atrium.

Endometrioid stromal sarcomas are rare malignant tumors which simulate uterine stromal sarcomas but occur in extra-uterine sites like the ovary, vagina, cervix, rectovaginal septum, parametrium and fallopian tube as well as extra-gonadal sites; including the gastrointestinal tract, omentum, mesentery, liver and sciatic nerve [1]. These tumors constitute 0.2% of uterine malignancies and occur over a wide age range of 11–76 years [2]. The pathogenesis of such extra-uterine sites is unclear; however, there is a frequent association between such sites and endometriosis, raising the possibility of malignant transformation of endometric foci [3]. In this context, we report a case of an ovarian endometrioid stromal sarcoma removed from the heart which was first diagnosed and treated as a thrombus emphasizing the importance of taking the consideration of a tumor mass in the differential diagnosis of an intra-cardiac thrombus.

Case report

A 30-year-old female patient was referred to our University hospital for removal of what was thought to be a thrombus in the inferior vena cava and right atrium. 2 months before referral, the patient developed abdominal discomfort and swelling. Abdominal ultrasound showed hepatomegaly and ascites as well as obliteration of the inferior vena cava with a thrombus. Echocardiography revealed a mass in the supra-diaphragmatic IVC as well as right atrium which was interpreted as a thrombus. CT chest and abdomen showed the same picture with no evidence of pulmonary emboli. The patient was anti-coagulated and a cause of a hypercoagulable state was searched for, but all investigations were negative. There was a positive history of nulliparity due to endometriosis followed by the appearance of a low-grade malignant right ovarian tumor 4 years ago. The tumor was removed through a unilateral salpingo-ovariectomy followed by chemotherapy due to the patient's wish to have children.

One month later, the abdominal distension resolved but the patient developed, despite anticoagulation, edema of the

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right lower limb followed by the left one. Lower limb venous Doppler study detected bilateral ilio-femoral deep venous thrombosis. Abdominal ultrasound and CT abdomen were repeated and showed extension of the thrombus down to the femoral veins with partial recanalisation of the proximal IVC draining the hepatic and renal veins. At that point, the patient was referred to our surgical department due to propagation of the thrombosis (Fig. 1).

Through a median sternotomy and after full heparinization, cardiopulmonary bypass (CPB) was established by cannulating the ascending aorta and superior vena cava (SVC). Due to the extensive bilateral ilio-femoral thrombosis, cannulation of a femoral vein or IVC was not attempted. The patient was cooled down to a core temperature of 22 °C. The heart fibrillated at a temperature of 28 °C so an aortic cross-clamp was applied and cold blood antegrade cardioplegia was given as myocardial protection. Upon reaching the target temperature of 22 °C, total circulatory arrest (TCA) was initiated and the right atrium was opened. A firm yellowish lobulated mass 3 × 4 cm was found originating from the IVC and extending to the right atrium. The mass was removed in toto with its small-sized attachment to the IVC endothelium. The IVC was freed from remnants of an organized thrombus on its inner wall and another cannula was inserted into the IVC before restoration of the circulation. The period of TCA lasted for 37 min during which retrograde cerebral perfusion through the SVC cannula was performed. The patient recovered rapidly and showed an unremarkable postoperative course (Fig. 2).

Histological examination of the mass revealed malignant spindle cell cells with numerous mitotic figures and increased stromal vasculature (Fig. 2).

Upon immunohistochemistry, the strong positive reactivity for vimentin and cluster of differentiation 10 (CD10) as well as the negativity for Wilms tumor 1 (WT1), inhibin, desmin and epithelial membrane antigen (EMA) suggested a stromal tumor of mullerian duct cell line. Tumor cells were positive for progesterone and estrogen receptors suggesting to a tumor of the gynecologic origin. Referring to the ovarian tumor excised 4 years before the diagnosis, a low-grade endometrioid sarcoma was confirmed. A whole body PET/CT scan was performed and showed no metabolically active 'bright' spots in the uterus, the other ovary or other organs that could be interpreted as malignancy. The patient was referred to an oncologist to receive further chemotherapy.

Discussion

Despite being considered a benign lesion, endometriosis may be associated with some malignant criteria like the ability to express growth factors for proliferation and neoangiogenesis, susceptibility of relapse after excision and ability to invade adjacent and distant tissues [2, 3]. In addition, although the fact that endometrioid stromal sarcomas have better prognosis than other sarcomas [4], yet relapse (whether local or distant) may occur in up to 56% of cases, even as late as 20 years after surgery [5]. Our case gives a history of low-grade ovarian endometrioid stromal sarcoma 4 years before the reappearance of the same tumor in the IVC and right atrium. To our knowledge, this is the first time to report an ovarian endometrioid stromal sarcoma in the IVC and right atrium which could be a metastasis from the

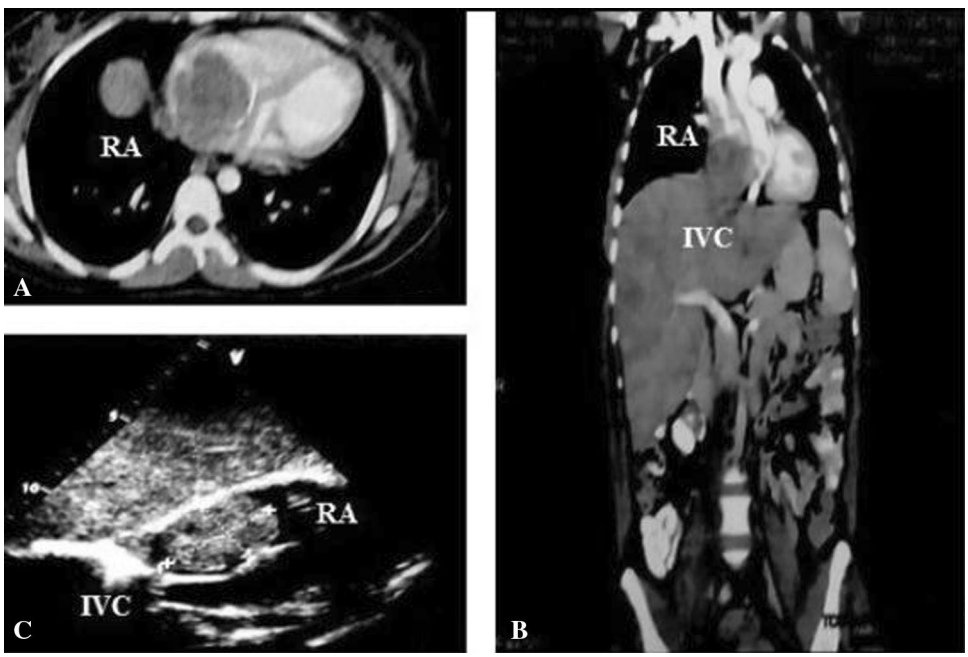

Fig. 1 **a** Sagittal section of CT chest showing the right atrium (RA) occupied by the mass. **b** Coronal section of the CT chest showing the mass in RA and the inferior vena cava (IVC). **c** Echocardiography confirming the mass in RA and IVC

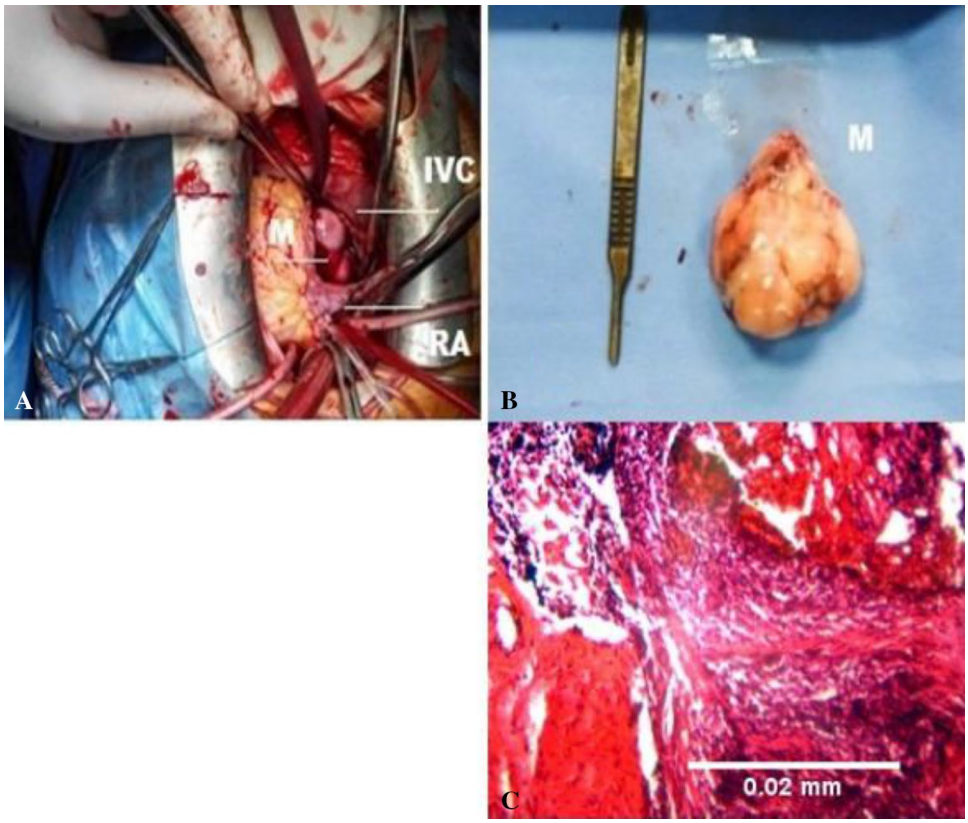

Fig. 2 **a** Intraoperative photo showing the mass (M) occupying the right atrium (RA) and inferior vena cava (IVC). **b** Intraoperative photo showing the removed mass. **c** Microscopic picture showing spindle cells with numerous mitotic figures and increased stromal vasculature

primary ovarian sarcoma or less likely a new transformation of an endometric focus.

Many tumors in the right atrium and IVC are initially interpreted as an extensive thrombus due to rarity of such tumors and associated thrombus formation distal to the tumor as a result of blood stagnation [6]. In addition, differentiation between a thrombus and a mass according to their enhancement characteristics upon imaging modalities such as CT and MRI is sometimes unreliable and accurate diagnosis can be only made by means of histological evaluation [7]. Therefore, a high suspicious index in the differential diagnosis due to the vague clinical presentation is required and a multidisciplinary team of a cardiac surgeon, internist, radiologist as well as oncologist should be involved (not only anticoagulation).

If a mass is suspected, an open approach using CPB should be considered. In this context, a hypothermic circulatory arrest may be required to offer a bloodless field for tumor resection [8].

Conclusion

Entometrioid stromal sarcomas are seen in extra-uterine as well as extra-gonadal sites and have a strong association with endometriosis. A mass should be always considered in the differential diagnosis of an extensive thrombosis of the venous system reaching the IVC and right atrium and its removal with the help of CPB should be attempted.

Compliance with ethical standards

Conflict of interest The authors have no conflict of interest to declare.

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