CASE REPORT

A big blob in the right atrium representing a tumour thrombus extending from uterine fibroids

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A 33-year-old woman presented with a history of lower abdominal pain, poly-menorrhagia, nausea and some weight loss in the last 2 months. Her blood investigations were unremarkable. However, the computed tomography of abdomen and pelvis showed a large mass arising from uterus highly suggestive of a large uterine fibroid. The tumor has invaded the iliac veins and the tumor thrombus has propagated all along inferior vena cava and extended to right atrium. The echocardiography showed a large multilobulated free floating mass in right atrium and prolapsing into right ventricle. The stalk of the mass was attached with tumor thrombus in inferior vena cava. The patient was started on anticoagulation and advised to do tissue biopsy and surgical resection of the tumor.

Background:

Right atrial mass is not an uncommon condition. Atrial myxoma is the most common cause of RA mass. The other important differential diagnoses are thrombus and metastatic tumor including tumor thrombus. Tumour thrombus is the extension of an invasive mass originating from the primary lesion to spread through inferior vena cava and reaching the right heart cavity. Tumor thrombus is a rare condition and caused by tumor of kidney, uterus, adrenal and liver. It is diagnosed by echocardiography and confirmed by tissue biopsy. Computed tomography and positron emission tomography also helps in the diagnosis. They are treated by surgical resection and in few conditions; it can be treated by chemotherapy or radiotherapy depending on the type of the tumor.

Case presentation:

A 33 year old young married lady presented with history of lower abdominal pain, polymenorrhagia, nausea and some weight loss for the last few months. She denied any history of chest pain, shortness of breath, palpitation, syncope or swelling of feet. There was no history of use of oral contraceptive. She had surgical history of bilateral oophorectomy 6 years ago but no details were available.

On examination her vital signs were normal. In cardiovascular system the heart sounds were audible and there was 3rd heart sound, plop. Her chest was clear. She has palpable mass in the lower abdomen.

Investigations and evaluation:

The electrocardiogram and X-ray chest were normal. Her initial blood investigation showed haemoglobin of 17.3 g/dl, white blood count of 13.7 X 103 /uL, platelet count of 83 X 103 /uL, random glucose of 160 mg/dl, creatinine of 0.9 mg/dl, amylase of 18 U/L, lipase of 9 ul, direct bilirubin of 1.1 mg/dl, indirect bilirubin of 0.6 mg/dl,

total bilirubin of 1.7 mg/dl, calcium of 9.3 mg/dl, dimer test of 1.53 mg/dl, erythrocyte sedimentation rate of 12 mm/1hour, fibrinogen of 973 mg/dl, prothrombin time of 13.1, partial thromboplastin time of 44.4, international normalized ratio of 1.02, sodium of 135 mmol/l, potassium of 3.9 mmol/l, procalcitonin of 35.65 ng/ml, albumin of 4 g/dl, alkaline phosphatase of 43 U/L, alanine transaminase of 18 U/L, total protein of 7.1 g/dl.

Computed tomography with contrast of abdomen and pelvis reported a huge uterine mass with lobulated contour and heterogenous enhancement, most likely fibroid, in the lower abdomen. It measures 24 x 2 x 15.5cm displacing bowel loops and urinary bladder with no evidence of obstruction or invasion (figure 1).



Figure1: Computed Tomography of abdomen showing large uterine mass (black arrow).

There is persistent hypodense filling defect seen in right iliac vein and inferior vena cava (IVC) extending up to right atrial (RA) level (figure 2). The texture and contrast enhancement are like that of uterine mass is highly suggestive of tumor thrombus.



Figure 2: Computed Tomography of abdomen showing long tumor thrombus in inferior vena cava (IVC) (follow black arrows).

Transthoracic echocardiography showed normal size cardiac chambers and normal ventricular function. There was grade 2/4 tricuspid regurgitation and right ventricular systolic pressure was 40mmHg.



Figure 3: Echocardiography apical 4 chamber view: large lobulated mass in right atrium prolapsing into right ventricle (white arrows).

There was a large mutilobulated mass of 6.2 x 2.6 cm seen in right atrium, freely mobile and prolapsing into right ventricle in diastole (figure 3). The stalk of the mass is attached to tumor thrombus in IVC (figure 4).



Figure 4: Echocardiography in subcostal view showing stalk of the RA mass attached with tumor thrombus in IVC (white arrows).

Most likely the mass is a tumor thrombus extending from the uterine tumor all the way through IVC to right atrium.

Differential diagnoses:

1- Tumor thombus extending into the right heart cavity.
2- Right atrial myxoma.
3- Thrombus in the right atrium.
4- Vegetation in right atrium
5- Hydatid cyst in the right atrium.
6- Metastatic tumor in the heart.

Management and follow up:

The patient was treated initially conservatively and anticoagulation with warfarin was started since the tissue diagnosis is not yet done. She was advised to do biopsy of the mass and would require surgical resection. Patient preferred to go back to her country to do further management. Patient went back to her country for surgery but she lost to follow up.

Discussion:

Cardiac tumours can be primary or secondary. The metastatic tumors are far more common than primary ones. The most common secondary tumors that metastasize to heart are carcinoma of breast, lung, esophagus, leukemia and malignant melanoma. The tumours can invade heart by any of the following path either by direct infiltration, through blood and lymphatic, or intraluminal venous extension [1].

Tumour thrombus is the term given to an extended invasive mass originating from the primary lesion to the right atrium or right ventricular cavity. The tumors that spread by invasion IVC and reaching to right atrium causing intra-cavitary mass include carcinoma of kidney, hepatocellular carcinoma (HCC), uterine tumors like leiomyoma and adrenal carcinoma [1]. The most common tumor invading inferior vena

cava and spreading to right atrial cavity is renal cell carcinoma. The rate of such extension ranges between 4 to 25% [2].

The history of tumour thrombus is dated back to 1977 when Kerber RE and his colleagues reported an angiographically diagnosed case of a tumour thrombus originating from hypernephroma in a 61-year-old woman [3]. The pathogenesis is postulated as the tumour cells invade the vascular cavity in the inferior vena cava and then coagulation cascade build on top of it with fibrin deposition and extension of the thrombus.

The literature does not have any large studies on tumor thrombus but there are many case reports and few small case series. The extension of the tumour thrombus into the right atrium mimics a right atrial myxoma which is the most common primary benign cardiac tumor.

In a small case series of 14 patients, by Locali RF et al, who underwent surgical resection of the tumorthrombus. The underlying primary tumors were Wilm'stumor, clear cell carcinoma of kidney and adenocarcinoma of adrenal gland. All the patients had tumor extension in IVC and 62.4% among them developed RA invasion [4].

Panday H, published a rare case of hepatocellular carcinoma (HCC) having extension of tumor in portal vein that spreaded into IVC and reaching up to right atrium. The patient did not have any cardiorespiratory symptom [5].

A similar case cited by Bennett ES et al, in a 58-year-old woman who was sent for evaluation of a systolic murmur. Her echocardiography showed large lobulated mass extending from IVC into RA and crossing tricuspid valve and reaching up to right ventricular outflow tract. She was thoroughly investigated and found to have leiomyoma of uterus and the biopsy of the tumor thrombus confirmed the diagnosis [6].

The tumor thrombus is usually suspected on echocardiography and confirmed by the tissue biopsy of the mass. However, Davidson T in original paper mentioned that Fluorine-18 fluorodeoxyglucose (F-FDG) positron emission tomography (PET)/Computerized tomography (CT) may help to diagnose tumour thrombus and to differentiate from venous thrombus. This may help in the management plan before tissue diagnosis [7].

The treatment of a tumour thrombus and its primary site is surgical excision along with chemotherapy and/or radiotherapy. In another small case series by Tsuji Y et.al who did surgical resection of the tumour thrombus and given chemotherapy for the primary tumor. He found favourable outcome with such therapy [8]. He concluded radical resection with chemotherapy for renal tumors gives palliation and may cure the renal tumors extending to the heart.

According to the type of underling primary tumour the initial chemotherapy or radiotherapy can be used in the treatment of the tumour thrombus. Oberholzer HF, et al treated 4 patients of tumour thrombus with chemotherapy secondary to nephroblastoma and all the patients showed favourable outcome [9].

Zeng ZC et al in his large comparative study of 158 patients with hepatocellular carcinoma with invasion of IVC with tumor thrombus were treated with external beam radiation therapy (EBRT). He found that patients treated with EBRT had prolonged

survival as compared to those who received only with conventional treatment [2]. Moreover, Lee DS et al in published article mentioned the combination therapy with transarterial chemoembolization (TACE) and EBRT is found to be more effective in prolonging survival and controlling IVC tumour thrombi and in patients with HCC [10].

In our case the patient found with a huge uterine fibroid with extension of the tumour thrombus into the IVC and creping to reach the right atrium. Biopsy of the uterine mass and the tumor thrombus in IVC is required to confirm the diagnosis. Unfortunately, the patient got discharged herself and would like to do further treatment in her own country.

Learning and take home messages:

- Free floating right atrial thrombus is life threating condition which can complicate to pulmonary embolism or can cause tricuspid valve obstruction.
- It can be associated with deep vein thrombosis or malignancy.
- Surgical resection, chemotherapy and radiotherapy are the mode of treatment depending on the type of the tumor.
- Radiological characteristics of the tumour thrombus may alleviate unnecessary use of anticoagulation.

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