Renal Angiomyolipoma – Sarcomatous Transformation and Cavoatrial Extension

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Introduction

Renal Angiomyolipoma occurs both sporadically and in association with tuberous sclerosis [1]. Earlier regarded as a hamartoma, angiomyolipoma is now believed to be neoplastic, showing clonal non random inactivation of the X chromosome [2]. Though generally regarded as benign, angiomyolipomas rarely produce complicated clinical courses that can be life threatening. It is a common cause of severe perirenal and retroperitoneal haemorrhage [3]. More rarely they display malignant potential with rapid growth and metastasis. These malignancies associated with angiomyolipoma include malignant epithelioid angiomyolipomas and sarcomatous transformation of one of the components of the tumor, most commonly leiomyosarcoma [3–6]. Extrarenal growth and vascular invasion are rare [7].

Case Report

A 55 year old lady presented with retrosternal discomfort for one year, exertional dyspnoea and easy fatigability for 6 months. There was no family history of tuberous sclerosis. General examination was normal. Cardiovascular examination

revealed loud first heart sound and precordial middiastolic murmur. Abdomen was clinically normal.

Laboratory investigations including haemoglobin, leukocyte count and renal function tests were normal. Electrocardiogram revealed left anterior hemiblock. Echocardiogram revealed an 8×4 cm right atrial mass extending into inferior vena cava [IVC]. IVC was dilated by tumor and was not collapsible. Left ventricular function was normal. Coronary angiogram was normal. Contrast enhanced computed tomogram [CECT] of the abdomen revealed a 10.2×11.4×16.2 cm mass arising from left kidney with dense inhomogeneous enhancement on post contrast study. The attenuation value of the mass was variable, ranging from –28 to +36 Hounsfield units in different areas of the mass. The tumor thrombus is seen extending into left renal vein, inferior vena cava, right atrium and projecting into right ventricle [Fig. 1]. Chest X ray was normal. CECT of the chest showed no evidence of lung metastases.

The patient was planned and taken up for surgery. Upon laparotomy, a 15×10 cm mass was noted in the left kidney with no other intraabdominal gross disease. After sternotomy and systemic heparinization, standard cardiopulmonary bypass was established with aortic and bicaval cannulation. Right atrium was opened through the auricular appendage and cavoatrial portion of the tumor was mobilized [Fig. 2]. Left renal vein was opened and tumor thrombus was fractured. The cavoatrial portion of tumor thrombus was extracted through the atriotomy and atrial opening was sutured closed. Left radical nephrectomy was performed.

Histopathology of the specimen revealed a spindle cell tumor with moderate pleomorphism [Fig. 3]. Tumor tissue was subjected to immunohistochemistry. It was positive for HMB 45, SMA, Caldesmon, Desmin, CK and EMA. Ki67 index was 30%. A diagnosis of renal angiomyolipoma with sarcomatous transformation [leiomyosarcoma] was made.

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Fig. 1 Left renal tumor with cavoatrial thrombus projecting into right ventricle

Fig. 2 Tumor thrombus visible after cardiopulmonary bypass and right atriotomy

Fig. 3 Light microscopy shows spindle cells along with adipocytes

Discussion

Renal angiomyolipoma usually has a benign course and treatment is based on lesion size and symptoms. Progressive enlargement and venous thrombosis are rare complications [6]. Diagnosis of sarcomatous transformation in angiomyolipoma is aided by immunohistochemistry. Malignancies associated with renal angiomyolipoma include malignant epithelioid angiomyolipoma and sarcomatous transformation of one of the components of the tumor, most commonly leiomyosarcoma. Other types of sarcomas that can occur in renal angiomyolipoma include liposarcoma and fibrosarcoma. Sarcomatous transformation in renal angiomyolipoma is exceedingly rare [8]. To the author's knowledge, there are 28 reported cases of malignant transformation of renal angiomyolipoma till date, including malignant epithelioid angiomyolipomas and sarcomas.

In conclusion, our case reminds us of the extremely rare possibility of sarcomatous transformation of renal angiomyolipoma, of its possibility to extend through renal vein and inferior vena cava to right atrium and feasibility of safe resection even with such extension of tumor thrombus.

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