Assistance by vena cava filter in treatment of renal angiomyolipoma with inferior vena cava embolus（two cases report and literature review）

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Abstract: Benign renal angiomyolipoma (AML) rarely presents with evidence of extension into the renal vein, inferior vena cava (IVC) or atrium. To explore the malignant characteristics and treatments of AML, two cases of benign renal AML with a tumor embolus to the IVC were reported. Patients were received right radical nephrectomy with IVC tumor thrombectomy, after vena cava filter was placed in IVC to avoid tumor emboli into atrium under interventional surveillance. Angiomyolipomas were testified by pathology. Patients were totally asymptomatic after operation, and post-operatively review showed no evidence of recurrence and metastasis. Benign renal AML may invade the renal vein and inferior vena cava, showing malignant characteristics. In view risk of tumor embolus into heart and lung, surgical treatment of renal AML and inferior vena cava embolus should be recommended.
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Key words： renal angiomyolipoma；vena cava embolus；thrombectomy

1. Introduction

Renal angiomyolipoma (Angiomyolipoma, AML) is the most common mesenchymal renal neoplasm, and is belong to the classification of perivascular epithelioid cell tumors, also referred to as "PEComas". Molecular markers of melanoma and smooth muscle are co-expressed in these tissue,such as HMB-45 (human melanoma black-45)、 Melan-A (Human Melanoma Marker-A) and SMA(smooth muscle actin) (Martignoni et al., 2008). The typical renal AML is derived from the mature development of abnormal renal differentiation of mesenchymal tissue, and consist of benign proliferation of vascular, smooth muscle and adipose tissue in different proportions. There were fewer cases reported that renal AML invaded into the inferior vena cava and regional lymph nodes. Here, we report two cases of patients with right renal AML invaded into renal vein and inferior vena cava, and have successful surgery.

2. Material and Methods

2.1 Clinical data

Case 1：An 18-year-old female patient was diagnosed with the right kidney tumor in routine body examination for 1 year. The patient was in hospital with the chief complaint of intermittent right flank pain in recent 3 months without macroscopic and microscopic hematuria, and blood routine examination was normal. The ultrasound examination showed there was a 5.7cm × 4.5cm × 4.8cm hyperechoic renal hilum mass at the upper part of right kidney with inferior vena cava tumor thrombus. CT scanning showed there was a 6cm × 5cm hybrid density mass within the low-density area at the right kidney upper pole, and ipsilateral renal vein and inferior vena cava were filled with hypodense masses.it was considered the right renal vein and inferior vena cava angiomyolipoma. MRI (Figure 1) shown: On T2 weighted image with fat suppression, there was a heterogeneous low signal mass at right side pole of kidney and renal pelvis. The lesion was well defined, and expanded through right renal vein to inferior vena cava. There was filling defect in the inferior vena cava caused by the lesions. Primary diagnosis： 1. angiomyolipoma of right kidney 2.filling defect in right renal vein and inferior vena cava. Preoperative diagnosis: the right side of the renal angiomyolipoma with the right renal vein and inferior vena cava invasion.

Case 2: A 53-year-old male patient was in hospital with the chief complaint of right renal tumor diagnosed by body examination for fourteen days. There was no right lumbago, no macroscopic and microscopic hematuria on his body. The patient was diagnosed on hypertension for ten years in past medical history with the most hypertensive 160/100mmHg, and hypertension was well controlled by oral drugs. There were no similar renal lesion cases in his family. Various normal routine laboratory tests were all negative. The ultrasound examination showed there was a hyperechoic multiple occupation in the upper side of right kidney with the inferior vena cava hyperechoic mass. Its volume was estimated about 7cm×8cm×5cm. CT examination (Figure 2): the right

renal angiomyolipoma with the inferior vena cava fat emboli. Preoperative diagnosis: right renal angiomyolipoma with the right renal vein and inferior vena cava invasion.

2.2 Treatments

Before renal operation, Two cases were done IVC angiography through interventional fluoroscopy and vena cava filter (Aegisy XJLX3260, Shenzhen Pioneer Technology Company, China) was implanted by percutaneous shares venipuncture at vena cava of liver segment to prevent pulmonary embolism caused by tumor emboli during thrombectomy. Then, radical resection of right kidney and thrombectomy at inferior vena cava were done for each case.

3. Results

Case one:

Gross specimen: on the resected right kidney, it could be seen a yellow soft tissue tumor in the upper part and the renal hilum had been invaded by the tumor. A fat-like tumor thrombus was visible in the inferior vena cava, and thrombus was about 2 cm long. Mixed adipose tissue, smooth muscle tissue and blood vessels were visible on the microscopic specimen of the tumor and HMB-45, SMA, and Melan-A were positive by immunohistochemical staining. There were no tumor cells on all margins, including the ureter and tumor vascular. Diagnosis of renal AML with inferior vena cava emboli was confirmed by pathology. The patient had been totally recovered after operation. During one-year follow-up, no inferior vena cava tumor had been recurrence by CT scanning.

Case two:

Gross specimen: on the resected right kidney, it could be seen renal upper pole had been replaced by yellow soft tissue tumor and surrounded with small multiple yellow tumor. The renal hilum had been invased by the tumor. A fat-like tumor thrombus was visible in the inferior vena cava, and thrombus was about 4 cm long. A large number of mature adipose tissue and irregular vascular smooth muscle tissue were visible on the microscopic specimen of the tumor and HMB-45, SMA, Melan-A were positive by immunohistochemical staining. During two-year follow-up, no tumor had been recurrence in surgical region and inferior vena cava by CT scanning.

4. Discussions

AMLs are solid tumors and composed by the thick-walled blood vessels, smooth muscle cells and adipose tissue by different proportions of. Renal AML belongs to benign lesions of the kidney, representing 3% of all kidney solid tumors. 80% of AMLs are sporadic cases, while other 20% of AML patients are tuberous sclerosis. The incidence in patients without tuberous sclerosis is 0.1% for men and 0.2% for women. It is rare in children (Rakowski et al., 2006). Typical AML is slow-growing and local development. Accelerated growth of AML can be seen in larger tumors, multiple tumors and pregnancy.

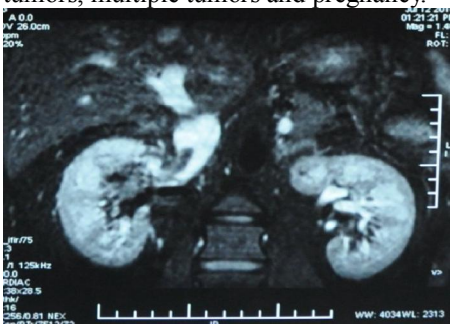

Figure 1: case one MRI(T2 weighted image with fat suppression): 1. angiomyolipoma of right kidney 2.filling defect in right renal vein and inferior vena cava.

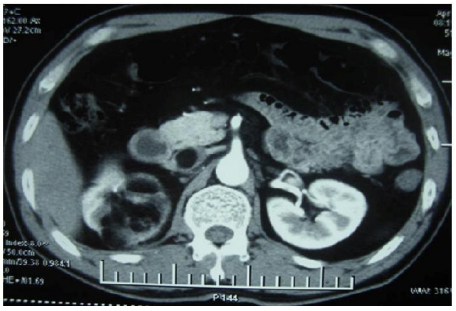

Figure 2: case two CT: the right renal angiomyolipoma with the inferior vena cava fat emboli.

In our study, tumor sizes of two cases were greater than 4cm, but there were no symptoms on patient's bodys. Usually majority of patients with renal AML less than 4cm are asymptomatic. Symptoms include fever, nausea, anemia, pain, a palpable mass, hematuria, hypertension and renal failure can be produced in patients with renal AML greater than 4cm tumor. When spontaneous bleeding happens in the huge AML, shock will be caused.

In our study, malignant tumor's characteristic to invade to the vena cava of is indicated in the two benign AMLs. There are several literatures to report malignant characteristics of AMLs, such as invasion to the vena cava and progress to the regional lymph nodes.Tan et al(Tan et al., 2010) reported the first case of AML-related pulmonary embolism. In their reviewed 35 cases of AML with venous invasion, only one case invaded to the renal vein, and 30 cases invaded to inferior vena cava and four cases had extensive involvement up to the right atrium. They also regarded that the appearance of either fatty thrombus in the IVC or the fatty infiltration of renal hilar lymph nodes in AML could not be considered as an characteristic of malignancy, but only of local aggressive behavior.

In a retrospective analysis of AMLs, Eble (Eble,

1998) found that more than 20 cases had invaded to vein and 39 cases had invaded to the regional lymph nodes, but all of the lesions were benign. After resection no case had progressed. Islam et al(Islam et al., 2004) found that there were two factors: 1 large volume 2 tumor located in the right side of the centerline. These two factors contribute to AML invasion to the renal vein and inferior vena cava. It is considered that the shorter and straighter course of the right renal vein contribute to the high frequent occurrence of tumors involving the vena cava. In addition, the large volume and multi-center tumor indicates that the tumor is easier to involve to the main vein. But the specific mechanism is unclear. Islam et al speculated the structures of the tumor vessels in AML were fragile, which might be related to the development of intravenous extensions of the tumor.

At present there are still controversial whether AML invasion to vena cava is benign or not. According pathology, it cannot be fully distinguished and predicted the biological behavior of the AML. Folpe et al (Folpe et al., 2005) observed a significant association between tumor size >5cm, infiltrative growth pattern, high nuclear grade, necrosis, and mitotic activity >1/50HPF and subsequent aggressive clinical behavior. It is considered potential malignancy as epithelial angiomyolipoma ,a rare subtype of AMLs, is diagnosed in histological pathological examination. Renal epithelioid AML (epithelioid angiomyolipoma, EAML) was reported involving of the inferior vena cava and regional lymph nodes(Kato et al., 2009). Luo et al(Luo et al., 2011) reported nine cases of renal EAML accompanied IVC invasion. EAMLs are malignant and invasive including biological recurrence, potentially lethal complications and metastasis.

With the increased understanding of AMLs, as well as the improvement of imaging diagnosis, accurate diagnosis of AML can help clinicians select appropriate treatment. For asymptomatic patients, surgical resection or close observation can be selected. Nephron-sparing treatment must be the first choice. Partial nephrectomy and arterial embolization are recommended for symptomatic AML or AML size greater than 4cm. When renal AML invade to the renal vein and inferior vena cava, it can result in lethal pulmonary embolism. Even in the absence of symptoms, it is an indication of surgical treatment. Radical nephrectomy and thrombectomy at vena cava are common treatments. Luo et al(Luo et al., 2011) reported that renal AML cases extended to vena cava, and eight of nine cases underwent radical nephrectomy and thrombectomy, and only one case accepted the treatment of partial nephrectomy. Yang et al(Yang et al., 2007) used interventional balloon catheter in the inferior vena cava to create a barrier to avoid cardiopulmonary embolism. Vena cava filter was utilized in our study by interventional manner to prevent intraoperative cardiopulmonary embolism and ensured the maintenance of circulating blood volume. Although the use of instruments are different, but the basic principle is similar to effectively reduce the surgical trauma.

From our AML cases, urologists should consider the rare possibility that renal AML can invade the renal vein and inferior vena cava. Considering the risk of potentially lethal cardiopulmonary embolism and death, radical nephrectomy plus tumor thrombectomy should be adopted in these AML cases even when asymptomatic.

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