CASE REPORT

Renal Angiomyolipoma with Inferior Vena Caval Involvement

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Summary

Renal angiomyolipomas are innocuous benign tumours which rarely behave aggressively. This is a case of a 48 year old Malay lady presenting with right sided abdominal pain associated with a large right sided abdominal mass. She was diagnosed with renal angiomyolipoma of the right kidney complicated by inferior vena caval tumour thrombosis. She successfully underwent a radical nephrectomy and inferior vena caval thrombectomy using cardio-pulmonary bypass and deep hypothermic circulatory arrest.

Key Words: Renal angiomyolipoma, Inferior vena caval thrombosis, Radical nephrectomy with inferior vena caval thrombectomy

Case report

A 48-year-old Malay lady presented with right iliac fossa pain for one week associated fever and vomiting. On examination, she had a tender palpable right lumbar mass. Laboratory investigations showed leucocytosis and microscopic hematuria. An ultra-sonogram revealed a 13 x 9cm hyperechoic mass involving the right kidney with mild hydronephrosis. Subsequent tomographic imaging (CT) confirmed the large hyperdense lesion involving the upper and middle poles of the right kidney and revealed an extension of the lesion into the inferior vena cava (IVC) via the right renal vein. These findings allowed the diagnosis of angiomyolipoma of the right kidney with IVC thrombosis to be made. To further delineate the tumour, magnetic resonance imaging (MRI) was carried out. The mass was hyperintense on T1-W1 series and hypointense on the fat suppressed sequence. The IVC thrombus was subhepatic extending to the point of the confluence of the hepatic veins. This was confirmed by an inferior vena cavogram.

She was initially treated with antibiotics and analgesia, following which her pain settled. She subsequently underwent a right radical nephrectomy with IVC thrombectomy using cardio-pulmonary bypass and deep hypothermic circulatory arrest. The procedure was performed one month after initial presentation. The tumour was approached with a bilateral subcostal incision. The right kidney was exposed and mobilised. Control of renal vessels was obtained and maintained with slings. Interaortocaval ligation of the right renal artery was carried out. Cardio-pulmonary bypass was then instituted via a median sternotomy. The right atrium was explored and noted to be free of thrombi. The patient was the heparinised and gradually cooled to a temperature of 18° Celsius. Once complete inactivity was confirmed by electro encephalogram, the patient was exsanguinated via the venous line. The right renal vein was divided flush with the IVC and the tumour thrombus was excised. The right kidney was thus excised en bloc. The IVC defect was then closed with non absorbable sutures. Slow rewarming then

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commenced, following which the patient was weaned off the by-pass. Total arrest time was eight minutes.

Post operatively the patient had intra-abdominal bleeding on the first post operative day and this necessitated a re-exploration. Bleeding from the cavotomy site and the adrenal bed was noted which was secured. Following this event the patient recovered without incident. On discharge she was well. She had suffered mild neuro-cognitive symptoms, which resolved spontaneously.

Histopathology showed a tumour measuring 15 x 10 x 5cm with a 9 x 1 x 1cm IVC extension. Microscopically, the tumour composed of an admixture of varying degrees of mature adipose tissue, bundles of smooth muscles and tortuous thick walled blood vessels. There was capsular and perinephric fat involvement but no features suggestive of renal cell carcinoma. Thus the diagnosis of renal angiomyolipoma with inferior vena caval thrombosis was made.

Discussion

Angiomyolipomas are hamartomatous lesions occurring most commonly in the kidney. Histologically, proliferation of smooth muscle, blood vessels and fat cells are the hallmarks of diagnosis. They are often incidental findings of routine ultrasonography of patients in their sixth or seventh decade. They can be confidently diagnosed with radiological imaging with characteristic sonographic, CT and MRI findings. However, occasionally they declare themselves in bizarre and alarming ways. These rare presentations include giant tumors presenting as painful, easily palpable renal masses and as causes of spontaneous retroperitoneal or intratumoral hemorrhage.

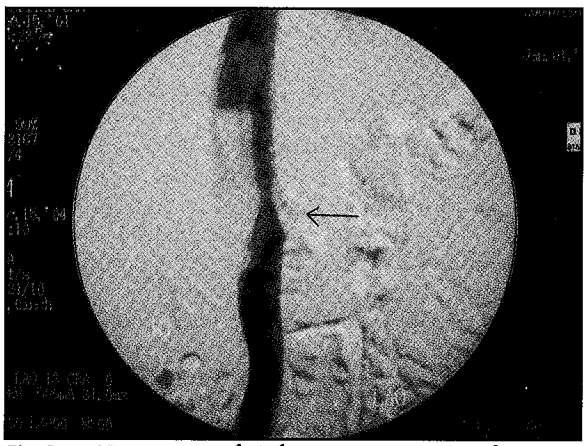

Fig 1 : Venogram of inferior vena cava showing tumor extension. Arrow marks the confluence of the IVC with the left renal vein.

Renal angiomyolipomas with inferior vena caval extensions are extremely rare. We are unaware of any previous reports locally. A.H. Manjurul Islam et al[1]. found that of the 26 reported cases, 81.5% arose from the right side. Eighty-one percent of the cases were women. The mean age of presentation was 46.1 years. In 63% of the cases the tumor arose from the upper pole and in 86.4% of the cases, the tumor occupied the central portion of the kidney. Average tumor size was 9.5cm in diameter. Thus, it was postulated that large angiomyolipomas, occupying the central portion of the right kidney were the contributing factors of inferior vena caval thrombus formation.

In this mode of presentation, the risk of fatal cardiopulmonary embolus and the suspicious behavior of the tumor necessitate radical nephrectomy and excision of the inferior vena caval thrombus. Other indications for nephrectomy for renal angiomyolipomas include suspicion of malignancy due to tumor behavior, control of retroperitoneal hemorrhage due to the tumor and giant angiomyolipomas replacing the entire kidney[2].

The use of cardio pulmonary bypass with a short period of circulatory arrest affords the surgeon a bloodless field during the excision of the tumor thrombus and this allows for satisfactory closure of the inferior vena caval defect[3]. Complications are mild provided arrest time does not exceed 30 minutes; these are in the form of slight cognitive symptoms such as temporary loss of recent memory or disorientation.

Further follow up post-operatively for these tumors may depend on the histology. Malignant transformation has been reported and is associated with large tumors, perivascular epitheloid cells and nuclear pleomorphism or atypia[4].

Conclusion

Large renal angiomyolipomas rarely extend into the renal vein, inferior vena cava or the right atrium. These are indications for radical nephrectomy with thrombectomy with or without cardio-pulmonary bypass. There are reported cases of malignant transformation of large renal angiomyolipomas and hence, post-operative surveillance is warranted.

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