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Aggressive Renal Angiomyolipoma with Renal Vein Thrombosis: A Rare Complication

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Abstract

Introduction: Renal angiomyolipoma (AML) is the most common type of benign mesenchymal tumour of the kidney. It is most often discovered incidentally on imaging, presented as a benign lesion without local invasion. Intravascular invasion to renal vein has been rarely reported in literature.

Case presentation: Here we report a case of renal AML with invasion into the renal vein in a 72 year-old man with hypertension presented with haematuria. Contrast Computed Tomography (CT) revealed a 6cm renal mass lesion with macroscopic fat at upper pole of the right kidney. There was accompanied fatty tumour thrombus invasion into the right renal vein. A positron Emission Tomography- Computed Tomography (PET-CT) scan showed the fatty renal mass was non-FDG avid. Patient had laparoscopic radical nephrectomy with pathology confirmed renal AML with vascular invasion.

Conclusion: Renal AML has the potential to extend into the renal vein, IVC, although it is classified as a benign tumour. Early imaging and therapeutic planning are necessary for the best outcome. Multi-disciplinary collaboration, including radiologist, urologist and the vascular surgeon, is highly recommended for optimal surgical treatment.

Keywords: Kidney; Angiomyolipoma; Renal vein; Thrombosis; Liposarcoma

Introduction

Renal angiomyolipoma (AML) is the most common type of benign mesenchymal tumour of the kidney. It is generally asymptomatic but can present with abdominal pain, haematuria, or bleeding. The involvement of the renal vein, inferior vena cava and regional lymph node has rarely been reported. In this study, we report a case of aggressive renal AML with tumour thrombus extension into the renal vein, presented with haematuria.

Case Presentation

A 72 year-old man, who had hypertension and impaired fasting glucose, presented with haematuria. He had no family history of tuberous sclerosis. He had no signs and symptoms of tuberous sclerosis. Apart from haematuria, he had no loin pain, shortness of breath or lower limb swelling.

Physical examination and blood test were unremarkable. A contrast Computed Tomography (CT) abdomen and pelvis was performed, which revealed a 6 cm renal mass lesion with macroscopic fat at upper pole of right kidney (Figure 1A-1C). It has a feeding artery from proximal anterior division of right renal artery (Figure 3). There was also fatty tumour thrombus invading into the right renal vein (Figure 2). There was no extension to inferior vena cava. There was no enlarged para-aortic lymph node.

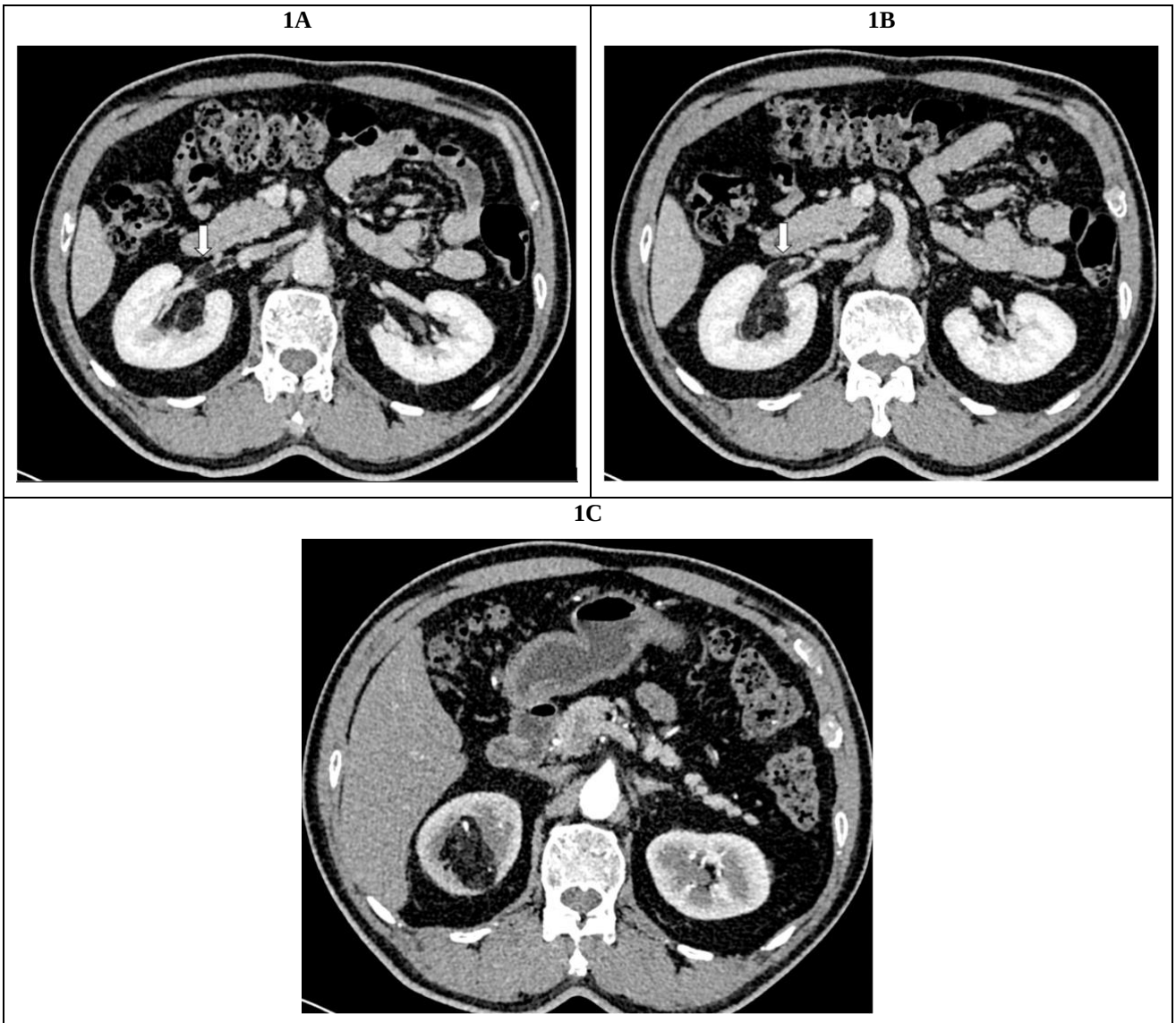

Figure 1: Contrast-enhancing axial CT images (Figure 1A-1C) show a renal mass at upper pole of right kidney containing fat attenuation with fatty tumour thrombus (arrow) invasion into right renal vein.

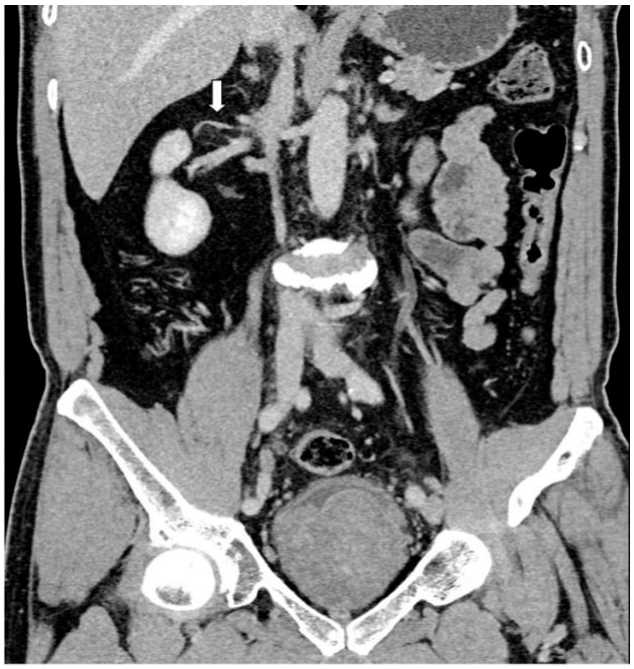

Figure 2: Contrast-enhanced coronal CT image shows fatty tumour thrombus (arrow) invasion into right renal vein.

Figure 3: Contrast-enhanced sagittal CT images show a feeding artery from right renal artery supplying to the fat containing renal mass at upper pole of right kidney.

The patient was referred to multi-disciplinary meeting for discussion. Abnormal aggressive feature of tumour extension into the right renal vein would be worrisome of other malignancy such as liposarcoma although renal angiomyolipoma (AML) with venous invasion has been reported in literature but exceedingly rare. Embolization was not the treatment of choice in this case. Further workup was suggested.

Patient then had a Positron Emission Tomography- Computed Tomography (PET-CT) scan for workup, which showed the fatty renal mass in the upper pole of right kidney was non-FDG avid (Figure 4A-4B). Fatty tumour thrombus invading to right renal vein was again seen. A diagnosis of renal angiomyolipoma was made.

Finally, patient had laparoscopic radical nephrectomy performed, with pathology result confirmed renal angiomyolipoma. Vascular invasion was present. Vascular and ureteric margins were clear.

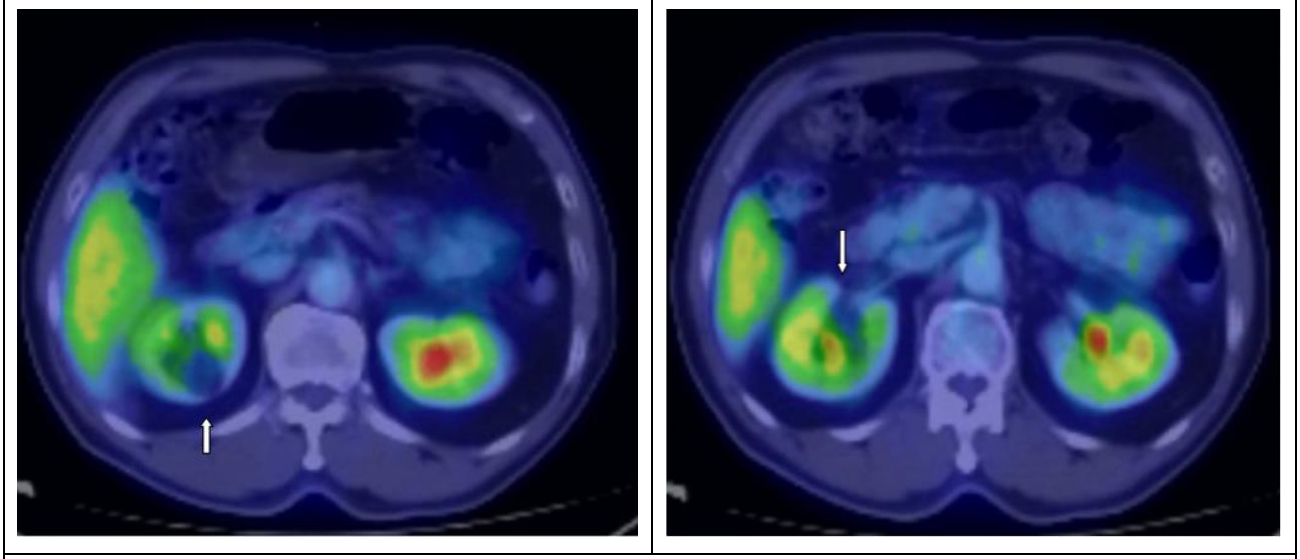

Figure 4: Positron Emission Tomogrpahy- Computed Tomography (PET-CT) (Figure 4A-4B) shows a macroscopic fat-containig renal mass (arrow) at upper pole of right kidney, which is non-FDG avid. Features are compatible with a renal AML. Liposarcoma is out of favour for the differential diagnosis.

Discussion

Renal angiomyolipoma (AML), is the most common mesenchymal tumour of the kidney comprised of mature adipose tissue, dysmorphic blood vessels, and smooth muscle [1,2]. This unusual benign tumour is found in 0.1% to 0.3% of the general population with 2 distinct groups: classic triphasic AML and monotypic epithelioid AML [1]. The mean age at presentation is 43 years, with a female predominance (male/ female ratio of 4:11) [3]. About 80% of AMLs are sporadic and the others associated with genetic syndrome such as tuberous sclerosis and lymphangioleiomyomatosis [4]. Most patients with sporadic AML are without symptoms, and AMLs are incidentally detected on cross-sectional abdominal imaging that is obtained for an unrelated reason [5].

Malignancy should be suspected on the basis of following criteria: presence of calcifications; large, irregular tumour invading the perirenal or sinus fat; large necrotic tumour with small foci of fat and association with non-fatty lymph nodes or venous invasion [6].

The indications for treatment, though somewhat controversial; the most studies recommend surgical treatment for large tumours (>=4 cm), even though they are benign [7,8]. Furthermore, AML invading the renal vein or IVC, irrespective of size, should be surgically removed even if it is asymptomatic [8,9]. Radiofrequency ablation or cryoablation may be effective for small, growing AMLs [10]. For patients with large tumours or a tumour thrombus in the renal vein and IVC, a nephron-sparing surgery or radical nephrectomy plus caval thrombectomy may be performed. For patients with acute, life-threatening hemorrhage, the preferred therapy is selective transarterial embolization [11].

The differential diagnosis of a fat-containing renal mass includes renal angiomyolipoma, perirenal liposarcoma, renal cell carcinoma (RCC). Perirenal liposarcoma and angiomyolipoma have different blood supply. Liposarcomas arise from the perinephric fat, which have tumour vessels extending from the fatty perirenal mass into the renal hilar vessel without traversing the parenchyma. Renal angiomyolipomas have blood supply and drainage from renal parenchyma [12]. Meanwhile, foci of fat and calcifications have been described in unusual cases of RCC.

To decrease the risk of pulmonary embolism in patients with a tumour embolus, some surgeons recommend implanting the IVC filter before nephrectomy [3]. Generally, a permanent IVC filter may cause several complications, including migration, thrombosis, filter fracture, IVC perforation and device infection [13]. A temporary IVC filter minimizes the risk of these complications and is easier to insert and retrieve. Thus, some paper recommended to place the filter before surgery to avoid these complications [14]. However, use of an IVC filter without prompt tumour thrombectomy is not recommended as to avoid ingrowth of the thrombus into the filter which can complicate filter removal.

Conclusion

Renal AML has the potential to extend into the renal vein, IVC, although it is classified as a benign tumour. Early imaging and therapeutic planning are necessary for the best outcome. Multi-disciplinary collaboration, including radiologist, urologist and the vascular surgeon, is highly recommended for optimal surgical treatment.

Radical nephrectomy with tumour thrombectomy is the most widely accepted surgical treatment following established principles of venous tumour thrombectomy. Nephron-sparing options may be considered in carefully selected patients depending on the primary tumour size and location.

Acknowledgements

Ethics approval

This study was conducted in accordance with the principles outlined in the Declaration of Helsinki. Verbal consent was obtained for the purpose of case study.

Author's Contributions

All authors contributed to the concept, acquisition and interpretation of data, drafting of the manuscript, and revision for important intellectual content. All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

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