Spontaneously ruptured renal angiomyolipoma with pseudoaneurysm – presenting as Wunderlich syndrome

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Patient: 28 years, male

Clinical History:

We report here an unusual case of a young patient presenting with hypovolaemic shock syndrome with severe right sided abdominal pain who underwent CT scan of abdomen along with colour Doppler ultrasound for evaluation of its cause.

Imaging Findings:

A 28 year old man, in a state of shock following sudden onset of severe right sided abdominal pain was admitted in our emergency department. There was no history of trauma. He was pale and drowsy with pulse rate of 124/min and non recordable blood pressure. After initial resuscitation with i.v. fluids, he was shifted to our radiology department where he underwent abdominal CT and colour Doppler sonography examinations. CT of the abdomen showed a large right perinephric haematoma displacing the kidney anterior with a heterogeneous size of 7.3x6.7cm containing few fat densities in the interpolar region (Fig. 1). There was evidence of intra-tumoural haematoma as well. Also a well defined rounded intensely contrast enhancing lesion of size 2x1.7 cm was seen in the mass (Fig. 2, 3). The provisional diagnosis of ruptured renal angiomyolipoma with pseudoaneurysm was made and for further confirmation of pseudoaneurysm, patient was screened with conventional and Colour Doppler sonography which showed a large echogenic mass at interpolar region of the right kidney with a hypoechoic rounded lesion in it (Fig. 4) showing characteristic to and from flow pattern in the neck (Fig. 5) that connected the pseudoaneurysm to the injured artery. Patient underwent partial nephrectomy with pathological confirmation of haemorrhagic angiomyolipoma with intratumoral pseudoaneurysm. The patient was well after surgery and at 3 month follow up.

Discussion:

Wunderlich in 1856 first described the clinical picture of spontaneous renal bleeding confined to subcapsular/ perinephric space in patients with no known underlying cause [1]. Patients may present with classic triad of symptoms- abdominal pain, a palpable mass and hypovolaemic shock as it was in our case. Angiomyolipomas are benign hamartomas composed of varying proportions of mature adipose tissue, smooth muscle cells, and blood vessels [2,3]. The abundant abnormal elastin poor vascular structures in AML make these lesions prone to aneurysm formation and rupture [1]. 80% of AML are sporadic with a female preponderance while 20% are associated with tuberous sclerosis. Sporadic AML are generally solitary and unilateral and occur more on the right side [1]. 15% of patients with renal AML present with haemorrhage [1]. Its most common clinical presentation is abdominal pain which usually occurs when it ruptures angiomyolipoma of greater than 4 cm in diameter are more likely to bleed [2,4]. Although intrarenal or perinephric haemorrhage is the usual complication of angiomyolipoma, pseudoaneurysm appears to be unusual. Intrarenal pseudoaneurysm is also a well known complication of
penetrating renal injury, renal surgery and percutaneous renal procedures [5,6]. Typical intrarenal AML may be diagnosed almost exclusively at imaging like sonography and CT. At sonography AML demonstrates marked increased echogenicity as was seen in our case. Occasionally other renal lesions like oncocytoma, cavernous hemangioma and malignancies like renal cell carcinoma and lymphoma may be indistinguishable from AML [7]. To more accurately establish the diagnosis, CT is most often used to detect foci of fat. However there are rare reports of intratumoral fat in renal cell carcinoma, oncocytoma and Wilms tumor [2]. AML with intratumoral haemorrhage are often difficult to characterize since intratumoral fat component can be obscured by blood [3]. CT is the method of choice for demonstration of perinephric haemorrhage with a sensitivity of 100% [1,8]. However for successfully identifying underlying renal lesion causing haemorrhage, its reported sensitivity is 57% [1]. For pseudoaneurysm, colour Doppler sonography patterns have been well described in the literature. It appears as anechoic to hypoechoic lesion that fills with colour signal on colour flow Doppler imaging with pulsed Doppler analysis at the level of neck shows to and fro pattern which signifies systolic feeding and diastolic draining arterial flow [5]. Once a patient is diagnosed with spontaneous perinephric haemorrhage due to angiomyolipoma with pseudoaneurysm, treatment options are either surgery or therapeutic embolisation [1]. Selective transluminal embolisation of pseudoaneurysm in this setting is an efficient treatment with lesions <4cm in size [5]. The benign nature of AML supports partial nephrectomy or other nephron sparing surgery [1]. Surgery also facilitates pathological confirmation. In conclusion, ruptured AML with pseudoaneurysm should be considered as a cause of Wunderlich syndrome. CT scanning is the method of choice for identifying perinephric haematoma and underlying cause being AML. However colour flow Doppler scanning appears to be the modality of choice for identifying intratumoral pseudoaneurysm that determines the risk of recurrent bleeding.

Differential Diagnosis List: Ruptured Renal Angiomyolipoma with Pseudoaneurysm formation.

Final Diagnosis: Ruptured Renal Angiomyolipoma with Pseudoaneurysm formation.

References:

Figure 1

Description: Contrast enhanced CT shows a heterogenous mass in interpolar region of right kidney with fat densities. Origin:
Figure 2

Description: Contrast enhanced CT shows an intensely contrast enhancing rounded intratumoral pseudoaneurysm. Origin:
Description: Coronal reconstructed CT image showing large renal mass containing fat densities with pseudoaneurysm with perirenal hematoma. Origin:
Description: Sonogram shows heterogeneously hyperechoic interpolar renal mass with a round hypoechoic intratumoral area, which shows bidirectional color flow signal on Doppler imaging. Origin:
**Description:** Color Doppler image shows characteristic to and fro flow pattern in spectrum obtained from neck of pseudoaneurysm. **Origin:**