

*Images in Nephrology*  
(Section Editor: G. H. Neild)

## Wunderlich's syndrome in spontaneous angiomyolipoma bleeding

Chun-Yu Lin, Yih-Huei Lin, Fu-Zong Wu and Huay-Ben Pan

Department of Radiology, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan.

Correspondence and offprint requests to: Huay-Ben Pan; E-mail: panhb@isca.vghks.gov.tw

**Keywords:** angimylipoma; wunderlich syndrome

A 57-year-old female presented to the emergency department with progressive right flank pain for 2 days. She denied recent traumatic events. Initial vital signs were stable with blood pressure 125/80 mmHg. Physical examination revealed right flank knocking tenderness. Laboratory data showed leukocytosis 20 180/ $\mu$ L and normal haemoglobin 10.5 g/dL. The routine urine analysis was normal. After clinical evaluation, an abdominal computer tomography (CT) revealed a huge fat-containing tumor with active contrast extravasation and hematoma formation over the right perirenal space (Figure 1A). A diagnosis of renal angiomyolipoma with spontaneous bleeding was made. Transarterial angiography of the right kidney was emergently performed with coil embolization of the hypervascular tumor with intralésional aneurysmal formations (Figure 1B). The patient's symptoms resolved shortly after embolization and she was scheduled for further renal-sparing surgery.

Renal angiomyolipoma are generally benign mesenchymal hamartoma composed of adipose tissue, tortuous blood vessels and smooth muscle. About 80% of renal angiomyolipoma occurs sporadically and 20% associated with tuberous sclerosis complex [1, 2]. Sporadic angiomyolipoma usually with a solitary nodule in middle-aged women; tuberous sclerosis complex with angiomyolipoma are usually bilateral and multiple, frequently with larger tumor size [2]. Clinical presentations of angiomyolipoma are usually subclinical until tumor size becomes progressively larger with mass effect or tumor rupture. Wunderlich's syndrome (spontaneous renal hemorrhage) is a rare but potentially life-threatening complication of renal angiomyolipoma in the setting of tumors >4 cm. Clinical manifestations usually include flank pain, a palpable mass, gross hematuria and hypovolemic shock. CT is a good choice of imaging modalities because it is sensitive to demonstrating a fat-containing renal mass with perirenal and retroperitoneal hemorrhage, which is characteristic of Wunderlich's syndrome due to spontaneous angiomyolipoma rupture. In addition,

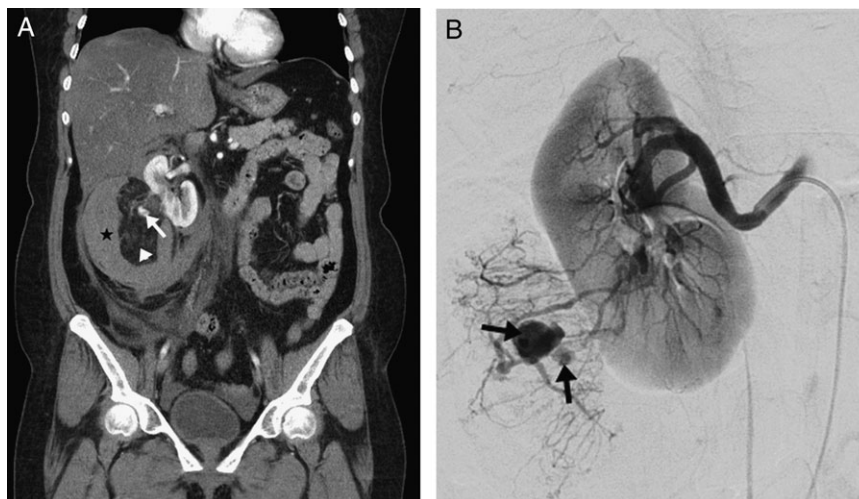


Fig. 1.

intralesional aneurysmal formations on the CT scan can lead to a greater possibility of tumor rupture [3].

Optimal treatment plans of angiomyolipoma depend on the tumor size and clinical manifestations. Conservative follow-up with imaging modality is recommended if tumor size is <4 cm in diameter. In acute tumor rupture, urgent transarterial superselective embolization can stop bleeding and save the life of patient. In symptomatic or larger angiomyolipoma with a high risk of bleeding, optimal treatment including elective preoperative transarterial superselective embolization prevention of and sequential renal-sparing surgery [4].

*Conflict of interest statement.* None declared.

## References

1. Carter TC, Angtuaco TL, Shah HR. US case of the day. Large, bilateral angiomyolipomas of the kidneys with tuberous sclerosis. *Radiographics* 1999; 19: 555–558
2. Blute ML, Malek RS, Segura JW. Angiomyolipoma: clinical metamorphosis and concerns. *J Urol* 1988; 139: 20–24
3. Albi G, Del Campo L, Tagarro D. Wunderlich's syndrome: causes, diagnosis and radiological management. *Clin Radiol* 2002; 57: 840–845
4. Williams JM, Racadio JM, Johnson ND *et al.* Embolization of renal angiomyolipomata in patients with tuberous sclerosis complex. *Am J Kidney Dis* 2006; 47: 95–102

*Received for publication: 03.01.11; Accepted in revised form: 14.01.11*