Netherlands
The Journal of Medicine

PHOTO QUIZ

A rare cause of haematuria

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CASE REPORT

A 36-year-old man with an unremarkable medical history presented to our emergency department with continuous lower abdominal pain radiating to the right side with fulminant macroscopic haematuria that started two hours earlier. Two weeks before admission the patient was treated with amoxicillin by the general practitioner for suspected pyelonephritis. Three days prior to admission the patient started using diclofenac because of increasing right-sided lumbar pain. There was no history of fever, chills, haematuria, pyuria or trauma. There was no positive family history of renal diseases. Physical examination revealed signs of shock (regular pulse 135 beats/minute; blood pressure 125/75 mmHg at presentation decreasing to 94/55 mmHg after one hour), with severe tenderness at the right costovertebral angle. Routine laboratory investigations showed anaemia (haemoglobin 5.5 mmol/l), serum creatinine 193 µmol/l, hyperkalaemia 5.3 mmol/l, lactate 5.2 mmol/l and a marked leucocytosis $36.2*10^9$/l. The fibrinogen, prothrombin time, partial thromboplastin time and thrombocyte count were all normal. The symptoms suggested intra-abdominal bleeding with compression of the ureteral structures. A contrast-enhanced computed tomography (CT) of the abdomen and pelvis was performed.

WHAT IS YOUR DIAGNOSIS?

See page 433 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 429)

A RARE CAUSE OF HAEMATURIA

DIAGNOSIS

Wunderlich's syndrome as a first manifestation of a renal angiomyolipoma

CT of the abdomen revealed massive (*figure 1, arrow*) active bleeding (*figure 2, arrow*) in the region of the right kidney. The kidney was enlarged, deformed and anteriorly displaced due to the haematoma with secondary upward displacement of the liver. The kidney showed a mass of approximately 9 x 9 x 9 cm with irregular contrast enhancement and focal black areas of fat as measured by their density.

The mass was likely to be an angiomyolipoma. A selective arterial embolisation of a side branch of the right renal artery was performed and stopped the renal bleeding. Recovery was uneventful and renal function normalised.

Severe acute subcapsular or perirenal bleeding was first clinically described by Carl August Wunderlich.[1] Wunderlich's syndrome is classically defined as non-traumatic spontaneous renal bleeding that can be caused by tumours, inflammatory vascular diseases, cysts, renal artery aneurysms, a renal vein thrombosis or arteriovenous malformations. However, the major cause of Wunderlich's syndrome is a renal angiomyolipoma.[2] Angiomyolipomas are rare, benign fat and myocyte-containing tumours often with a vulnerable vasculature found at approximately 0.3 to 2.1% of routine autopsies.[3] The chance of spontaneous rupture and bleeding increases in proportion to the size of the tumour and the degree of neovascularisation.[4] In general, close echographic follow-up or even prophylactic embolisation is advised in patients with an asymptomatic angiomyolipoma larger than 4 cm. The most common therapy for Wunderlich's syndrome is selective arterial embolisation, which often preserves the renal function. Unlike this case, these tumours are associated with tuberous sclerosis, which is present in approximately 10% of cases. Patients should be examined for characteristic skin lesions and other benign tumours (e.g. pulmonary lymphangioleiomyomatosis). Differentiation with renal epitheloid angiomyolipoma should be made as these tumours are associated with malignant degeneration. However, if intrarenal fat is found on CT, this rules out epitheloid angiomyolipoma.[5]

Figure 1. *Transverse CT scan of the abdomen*

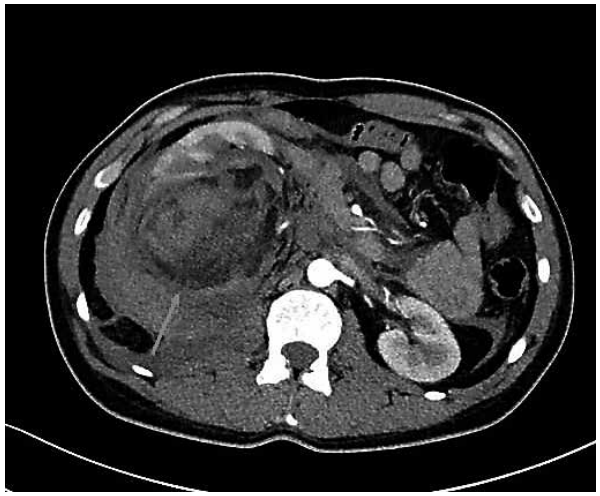
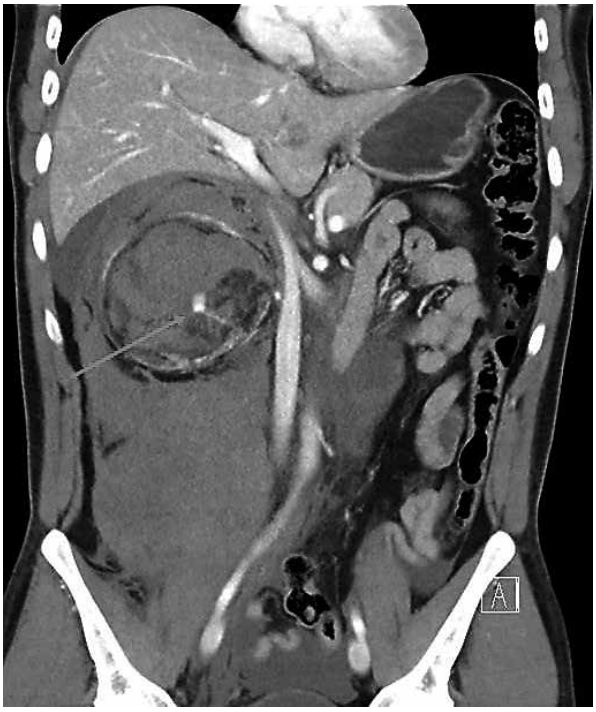


Figure 2. *Coronal CT scan of the abdomen*



REFERENCES

1. Wunderlich CA. Handbuch der Pathologie und Therapie. Ebner & Seubert: Stuttgart; 1852.
2. Albi G, del Campo L, Tagarro D. Wünderlich's syndrome: causes, diagnosis and radiological management. Clin Radiol. 2002;57:840-5.
3. Hajdu SI, Foote FW Jr. Angiomyolipoma of the kidney: report of 27 cases and review of the literature. J Urol. 1969;102:396-401.
4. Ploumidis A, Katafigiotis I, Thanou M, Bodozoglou N, Athanasiou L, Ploumidis A. Spontaneous Retroperitoneal Hemorrhage (Wunderlich Syndrome) due to Large Upper Pole Renal Angiomyolipoma: Does Robotic-Assisted Laparoscopic Partial Nephrectomy Have a Role in Primary Treatment? Case Rep Urol. 2013;2013:498694.
5. Tsai C-C, Wu W-J, Li C-C, Wang C-J, Wu C-H, Wu C-C. Epithelioid angiomyolipoma of the kidney mimicking renal cell carcinoma: a clinicopathologic analysis of cases and literature review. Kaohsiung J Med Sci. 2009;25:133-40.