

# A rare cause of spontaneous perirenal haemorrhage in a patient with ANCA-associated vasculitis

H.E. Boersma<sup>1</sup>, R.H.H. Nap<sup>1</sup>, W.P. Haanstra<sup>1</sup>, P.G.A. Hooijboer<sup>2</sup>, F.G.H. van der Kleij<sup>1\*</sup>

Departments of <sup>1</sup>Internal Medicine and <sup>2</sup>Radiology, Scheper Hospital Emmen, Emmen, the Netherlands, \*corresponding author: tel. +31 (0)591 691360, e-mail: fgh.vanderkleij@sze.nl

## CASE REPORT

A 51-year-old female presented with lower abdominal and flank pain. Her medical history revealed ANCA (PR-3) associated vasculitis with extracapillary necrotising glomerulonephritis that started 15 years ago. After several episodes of treatment and relapses, she developed end-stage renal failure four years later and started haemodialysis. Four years ago she had a spontaneous renal haemorrhage of her left contracted kidney that resolved with conservative therapy.

Physical examination showed a patient in circulatory shock with some tenderness in the lower abdomen. All laboratory results were normal except for a haemoglobin level of 3.4 mmol/l.

Ultrasonographic evaluation revealed a right-sided retroperitoneal haematoma. Rapid blood transfusion and fluid resuscitation was not sufficient to stabilise the patient. Therefore, emergency intervention angiography was performed to determine the source of the bleeding and to evaluate treatment options (*figure 1*).

**Figure 1.** Selective catheterisation of the right renal artery



## WHAT IS YOUR DIAGNOSIS?

See page 322 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 318)

A RARE CAUSE OF SPONTANEOUS PERIRENAL HAEMORRHAGE IN A PATIENT WITH ANCA-ASSOCIATED VASCULITIS

DIAGNOSIS

A diagnosis of multiple renal aneurysms (the several small black dots in the right kidney) was made based on the selective catheterisation of the right renal artery, with clear extravasation of contrast media from one of these aneurysms (right upper pole). Selective catheterisation of the involved segmental arteries was followed by successful obliteration by embolisation.

Spontaneous perirenal haemorrhage is a rare but dramatic clinical problem with different aetiologies.<sup>1,2</sup> The predisposing conditions responsible for this clinical entity include malignant and benign neoplasm, renal artery aneurysm, renal vein thrombosis, polycystic kidney disease, arteriovenous malformations and more rarely infectious disease and sickle-cell trait. Renal vasculitis is also mentioned as a cause, dominantly described in patients with classical polyarteriitis nodosa (PAN).<sup>1,2</sup> In PAN, small and medium sized arteries are involved. The pathological basis of aneurysm formation is active fibrinoid necrosis of the arterial media, followed by extension of the process into the intima and adventitia. Subsequently, an inflammatory response invades the layers of the vessel wall. If areas of segmental necrosis involve the elastic wall to a sufficient degree, an aneurysm may develop.

In most publications and textbooks, renal aneurysm formation has solely been attributed to PAN. Before the era of ANCA, in patients with suspicion of renal vasculitis, the absence or presence of renal aneurysms by diagnostic

angiography was even the main clue to discriminate between Wegener's granulomatosis and PAN, especially in cases where classical granuloma could not be detected. In previous reports, only two patients have been described with supposed Wegener's granulomatosis and spontaneous renal haemorrhage.<sup>1,3</sup> Only in one case was association with similar multiple renal aneurysm formation, as in this case, confirmed by angiography. This patient had the classical triad of biopsy-proven granulomas in the nose with pulmonary and renal involvement.<sup>3</sup> Therefore, this case appears to be the second case that shows us that renal aneurysm formation is not exclusively seen in PAN, but also in ANCA-associated vasculitis. In patients with ANCA-associated renal disease, we should be aware of renal aneurysm formation when spontaneous renal haemorrhage is detected.

REFERENCES

1. Daskalopoulos G, Karyotis I, Heretis I, Anezinis P, Mavromanolakis E, Delakas D. Spontaneous perirenal hemorrhage: A 10-years experience at our institution. *Int Urol Nephrol.* 2004;36:15-19
2. Reiter WJ, Haitel A, Heinz-Peer G, Pycha A, Marberger M. Spontaneous nontraumatic rupture of a contracted kidney with subcapsular and perirenal hematoma in a patient receiving chronic hemodialysis. *Urology.* 1997;50:781-3.
3. Baker, SB, Robinson DR. Unusual renal manifestation of Wegener's Granulomatosis. Report of two cases. *Am J Med.* 1978;64:883-9.