

Polyarteritis nodosa with multiple aneurysms and renal arteriovenous fistula successfully diagnosed by colour Doppler sonography

HongYan Wang · JianChu Li · YuXin Jiang · Qing Dai · Ying Jiang · Yong Hou · XiaoGuang Li

Received: 4 May 2010 / Accepted: 2 June 2010 / Published online: 18 June 2010
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Abstract Polyarteritis nodosa (PAN) is a vasculitis that typically affects small- and middle-sized arteries of multiple organs. The kidney is most commonly involved. To date, few cases of PAN involving the celiac artery and no cases of iliac artery or internal carotid artery involvement have been published. The most frequent lesions reported were due to occlusions and aneurysms. Finally, no cases of PAN causing arteriovenous fistula have been reported. Here we present a case of PAN with multiple aneurysms and renal arteriovenous fistula that was successfully diagnosed and followed-up by ultrasound. This case report describes the challenges in diagnosing PAN and highlights the importance of a holistic approach when encountering patients with multiple vascular diseases. Ultrasound should be considered the first-line approach in the diagnosis of PAN.

Keywords Polyarteritis nodosa · Ultrasonography · Vasculitis

HY. Wang · JC. Li · YX. Jiang (✉) · Q. Dai · Y. Jiang
Department of Diagnostic Ultrasound,
Peking Union Medical College Hospital,
Chinese Academy Medical Sciences,
N0.1 Shuai Fu Yuan, Dong Cheng District,
Beijing 100730, China
e-mail: yuxinjiangxh@126.com

Y. Hou
Department of Rheumatology and Immunology,
Peking Union Medical College Hospital,
Chinese Academy of Medical Sciences,
Beijing, China

XG. Li
Department of Radiology,
Peking Union Medical College Hospital,
Chinese Academy of Medical Sciences,
Beijing, China

Introduction

We present a case of polyarteritis nodosa (PAN) with multiple aneurysms and renal arteriovenous fistula that was successfully diagnosed and followed-up by ultrasound. The primary reasons a patient develops a renal arteriovenous fistula are congenital abnormality, trauma and renal biopsy. We report the first case of renal arteriovenous fistula caused by PAN. Ultrasound is less time consuming than angiography, is non-invasive and has few adverse effects. This procedure provides an interesting opportunity for further research.

Case report

A 44-year-old man, suffering from an intermittent fever of unknown origin accompanied by generalised weakness, pain in his lower right abdomen, vomiting and diarrhoea for 5 years, was admitted to the hospital 6 days after the above symptoms relapsed. In August 2004, he had fever, abdominal pain, weight loss of 15 kg in 2 months and blood pressure of 150/100 mmHg. He underwent an abdominal laparotomy for acute lower right abnormal pain in a local hospital. At the operation, necrosis, haemorrhaging and local inflammation were present in the patient's mesentery, and thrombi were widely spread in the mesenteric artery and vein. The patient did not feel well after the operation and was admitted to our hospital for further treatment. The patient had no family history regarding relevant vascular diseases, blood cholesterol was normal and did not smoke. Laboratory studies revealed an erythrocyte sedimentation rate (ESR) of 46 mm/h (normal 0–15 mm/h) and C-reactive protein (CRP) level of 11 mg/dl (normal 0–8 mg/dl). Autoantibodies, including rheumatoid factor, anticardiolipin antibody, antinuclear antibodies and

antineutrophil cytoplasmic antibodies, were negative. Blood cultures and hepatitis B surface antigens were negative. CT Angiography (CTA) and ultrasound showed the formation of aneurysms in the celiac trunk (Fig. 1), superior rectal artery and the left distant renal artery; superior mesenteric artery narrow. The right renal artery was normal; however, the bilateral internal iliac artery and left common iliac artery were fusiformly dilated. Together with the clinical, laboratory and ultrasonographic findings, the diagnosis was established as PAN. The patient was treated with oral prednisolone (50 mg/d) and intravenous cyclophosphamide (200 mg Qod), after which the patient's symptoms were alleviated. The dosage of prednisolone tapered and maintained at 10 mg Qod.

The patient was readmitted to our hospital in September 2008 due to symptom relapse. Ultrasound and CTA revealed an aneurysm formation in the right renal artery, an aneurysm reduction in the left renal artery, a fusiform dilation of the left internal carotid artery and the unchanged dilation of the bilateral iliac arteries. He was treated with larger dose of methylprednisolone (40 mg Q12 h intravenous) and cyclophosphamide sequentially. The patient's symptoms were alleviated.

In October 2009, the patient was admitted to our hospital to evaluate his condition. Physical examination revealed the patient was emaciated; a murmur was present on the upper and right flank of the abdomen. There was no enlargement of the superficial lymph nodes, liver or spleen. Investigation found that the ESR was 15 mm/h (normal 0–15 mm/h) and that the levels of CRP, creatine kinase and blood urea nitrogen were 8 mg/dl (normal 0–8 mg/dl), 124 $\mu\text{mol/L}$ (normal 53–132 $\mu\text{mol/L}$) and 7.54 mmol/L (normal 1.07–7.14 mmol/L), respectively. Midstream urine showed 3+ red blood cells.

Gray-scale sonography revealed right main renal artery and upper lobar branch irregular ectasia. Colour Doppler

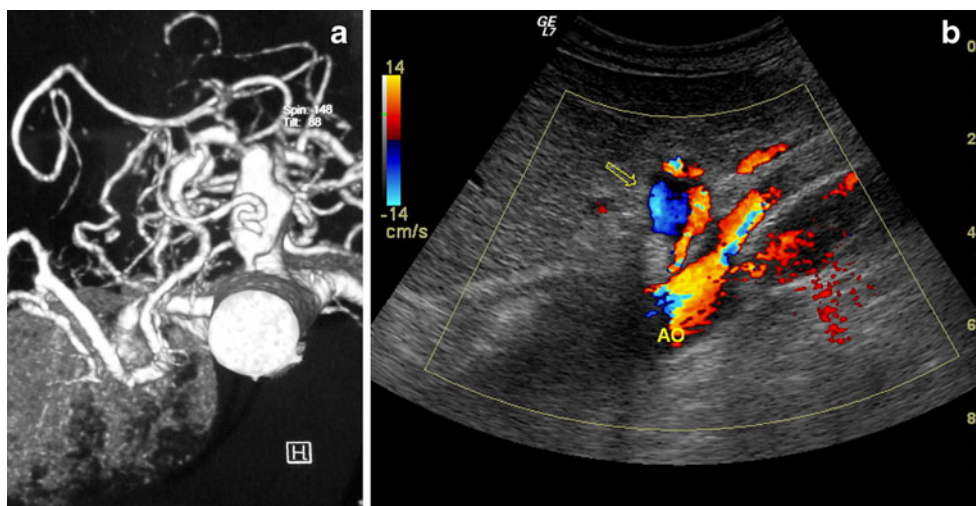
flow imaging clearly showed a communication between the right renal artery and vein. In addition, Doppler study showed an arterial waveform pattern with a very high peak systolic velocity (399 cm/s) and a very low resistant index (0.3) in the proximal right renal artery. The right renal veins and the inferior vena cava became markedly dilated to aneurysmal proportions where arterialisations of the venous waveform was detected (Fig. 2). Thus, renal arteriovenous fistula could be diagnosed by ultrasound. There were no significant changes of the multiple aneurysms identified by ultrasound follow-up. Angiography confirmed the diagnosis. At the time of detection, the size of the fistulas and the extent of arteriovenous shunting in the patient were significant. Thus, intervention was necessary to prevent complications, such as heart failure, bleeding or rupture. Angiographic embolisation was preferred to surgery because it is a procedure associated with low morbidity. The patient agreed to the angiogram embolisation of the fistula; however, the procedure was unsuccessful due to the migration of the coils inserted.

Discussion

PAN is a necrotising vasculitis involving medium and small arteries and sparing the venous system [1]. The characteristic pathological findings are fibrinoid necrotising inflammatory foci in the walls of small- and medium-sized arteries, with multiple small aneurysms. In the acute phase, the elastic lamina is destroyed, and the media undergoes fibrinoid necrosis. Microaneurysms arise from these weakened portions of the wall. In the reparative and chronic stages, fibroblast proliferation causes wall thickening and may produce areas of stenosis or occlusion [2].

The patient did not have oral ulcer and cardiovascular risk factors. He had increased inflammatory marker such as

Fig. 1 a In 2004, a 3D CTA showed an aneurysm of the celiac trunk and showed that the right renal artery was normal. **b** In 2004, ultrasound showed a proximal celiac trunk stenosis and distal celiac trunk aneurysm in which swirling flow was detected



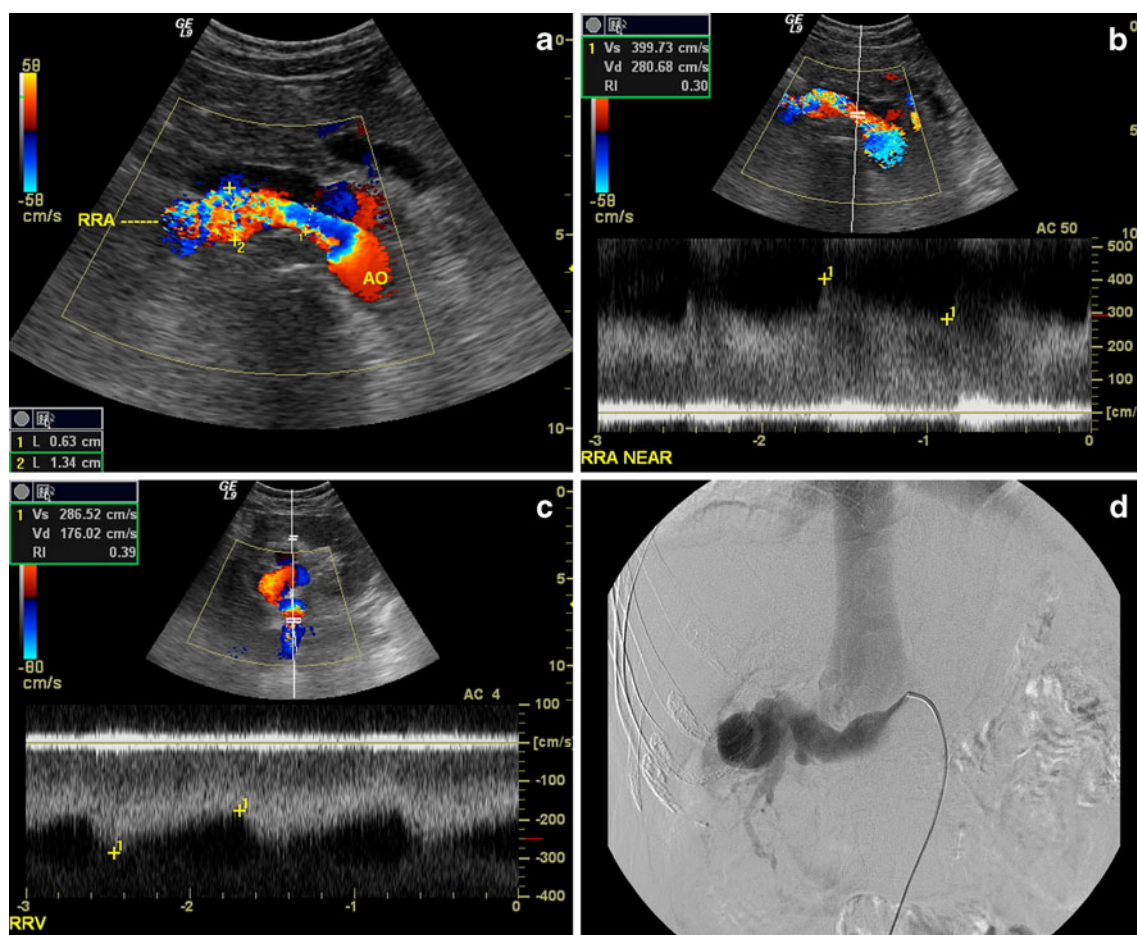


Fig. 2 The sonography and angiogram of the arteriovenous fistula in 2009. **a** Sonography showed the dilated distal right main renal artery, which had a diameter of 1.3 cm. The proximal right main renal artery was normal, with a diameter of 0.6 cm. **b** An arterial waveform pattern with a very high peak systolic velocity (399 cm/s) and a very low

resistant index (0.3) in the proximal right renal artery. **c** An arterial waveform pattern in the right renal vein. **d** A renal angiogram showing a large arteriovenous fistula with a lobulated aneurysm in the right kidney

ESR and CRP. He had multiple artery involvement. The symptoms responded to steroids and immunosuppressant treatment. So we think that the patient had primary vasculitis, and it is explored to further classify the vasculitis. According to the 1990 American College of Radiology criteria [3], the patient was diagnosed with PAN based on the findings of weight loss of at least 4 kg, diastolic blood pressure of >90 mm Hg, elevated blood urea nitrogen, weakness and arteriographic abnormality. PAN is a vasculitis that typically affects small- and middle-sized arteries of almost any organ. As seen in the spectrum of angiographic findings in PAN, the most commonly involved arteries were renal arteries [4]. To date, few cases of PAN involving the celiac artery have been published [5–10]. Moreover, no reports of PAN involving the iliac artery or the internal carotid artery have been published [5–12]. The most frequent lesions were due to occlusions and aneurysms [5]. Finally, no reports of PAN causing arteriovenous fistula have been published [5–13]. This study revealed multiple

aneurysms involving the renal, celiac, iliac and internal carotid arteries by angiography and ultrasonography. The findings of multiple small aneurysms on abdominal arteriograms have often been considered pathognomonic for PAN. It has been shown that angiography had a sensitivity of 89% and a specificity of 90% for PAN [6].

This case is presented as renal arteriovenous fistula caused by PAN. The primary reasons a patient develops a renal arteriovenous fistula are congenital abnormality, trauma and renal biopsy [12]. The patient had no history of trauma or renal biopsy. A congenital abnormality could be ruled out because his right renal artery was normal in the CTA taken 5 years before. Thus, we report here the first case, to our knowledge, of renal arteriovenous fistula caused by vasculitis. The kidney is a richly vascularised organ with small arteries and veins occurring close together [14]. A vasculitis lesion involving arteries and sparing the venous system may eventually become an arteriovenous fistula.

In a literature review, we found only two previous reports in which Doppler sonography was employed for diagnosis of PAN with arterial aneurysms [13, 15]. There were no cases of PAN causing arteriovenous fistula reported. Our case provides further evidence of the value of colour Doppler analysis in PAN patients. Colour Doppler imaging can quickly verify whether a lesion is indeed vascular, and the presence and size of a thrombus can be appreciated. Colour Doppler sonography can also evaluate complex flow patterns and thus help characterise and differentiate aneurysms from other vascular abnormalities [15]. The patterns of arteriovenous fistula using this technology include a communication between the right renal artery and vein, turbulent high-velocity flow present at the site of the fistula, low resistance flow present in the artery proximal to the fistula and artery-like flow detected in the vein [14]. This case could be diagnosed as renal arteriovenous fistula based on the ultrasound features below. A communication between the right renal artery and vein in the renal hilus was clearly visualised on colour flow imaging. Further examination detected a high velocity and a decreased resistance pattern from the proximal right renal artery, which was markedly enlarged. The right renal veins and the inferior vena cava became markedly dilated to aneurysmal proportions, where arterialisation of the venous waveform was detected. Dilation of the right renal veins and the inferior vena cava was thought to be caused by the jet from the fistula and the massive shunting of the blood from the right renal artery through the fistula to them. Colour Doppler patterns of abdominal aneurysms include dilation of the artery in which a mosaic colour configuration and bidirectional or swirling flow can be detected [14]. We successfully diagnosed and followed-up on the multiple aneurysms according to the ultrasound features.

Ultrasound is less time consuming than angiography, is non-invasive and has few adverse effects. Here, we show that it can identify aneurysms and arteriovenous fistula. CTA has the sensitivity to detect aneurysms in medium-sized arteries with a diameter as small as 3 mm [10, 11]. Ultrasound and CT, however, have the disadvantage of being incapable of clearly demonstrating very small aneurysms that can be seen only with conventional angiography [11]. The role of angiographic imaging in detecting microaneurysms remains to be shown [5, 6]. Angiography is still considered the gold standard for

imaging of PAN, but it is invasive. Even ultrasound techniques cannot replace the role of conventional angiography for diagnosis, but it can provide the first-line procedure for the follow-up in the future.

Disclosures None

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