Case Report

Acute Rupture of a Fibromuscular Dysplasia Related Renal Artery Aneurysm: Emergency Treatment with a Covered Stent

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We report the successful exclusion of a ruptured left renal artery aneurysm as a first presentation of fibromuscular dysplasia in a haemodynamically unstable 57-year-old man. The aneurysm was repaired in an emergency setting by deployment of a covered stent with a satisfactory result. Follow-up computed tomography confirmed successful exclusion of the aneurysm. A renal artery branch originating from the aneurismal sac was sacrificed with subsequent regional infarction. Our experience shows that the use of a covered stent is an effective, quick and life saving procedure in a ruptured renal artery aneurysm.

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Fibromuscular dysplasia (FMD) is a rare, non-inflammatory, non-atherosclerotic vascular disease that can affect almost any vessel bed. It is classified histologically into three main groups, according to the arterial layer most affected: intimal fibroplasia, medial dysplasia and adventitial fibroplasia, with medial dysplasia being the most common form.[1]

Renovascular FMD tends to affect primarily young and middle-aged women, and is often an incidental finding in imaging, manifested by a beaded pattern of the renal artery.[2] Renal artery aneurysms (RAA) may also occur in up to 10% of cases, with the risk of distal embolisation or rupture.[3,4] A ruptured RAA is an emergency situation with mortality rates that reach 80%.[5] We report a case of a ruptured RAA in a male patient that was treated successfully with the use of a covered stent.

Case presentation

A 57-year-old male non-smoker with a history of hypertension, hypercholesterolaemia and type II diabetes presented with sudden, severe, left-sided abdominal pain radiating to the back. Computed tomography angiography (CTA) performed in arterial phase with a bolus tracking technique revealed a 4.6 cm left renal artery aneurysm with stranding of the perinephric fat. A polar vessel originated from the aneurysm (Figure 1). Blood pressure was 100/63 mmHg and heart rate 118 beats per minute. On examination, the left renal angle was tender but the abdomen was soft. Twenty minutes after arrival the patient became haemodynamically unstable, blood pressure dropped to 86/58 mmHg and the patient was transferred to the interventional radiology suite for endovascular treatment. Under local anaesthesia, access from the right groin was obtained, and a 6 Fr 40 cm long sheath was placed in the origin of the left renal artery. Intra-arterial angiography confirmed the presence of the aneurysm (Figure 2a). The vessel originating from the aneurysm in the CT

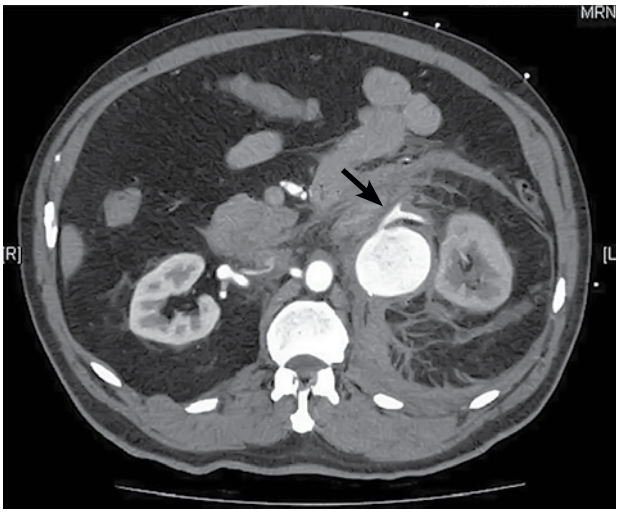

Figure 1. Computed tomography angiography in arterial phase showed a 4.6 cm left renal artery aneurysm with stranding of the perinephric fat. Twenty minutes later the patient was haemodynamically unstable. The branch that emerges from the aneurysm needed to be sacrificed (arrow).

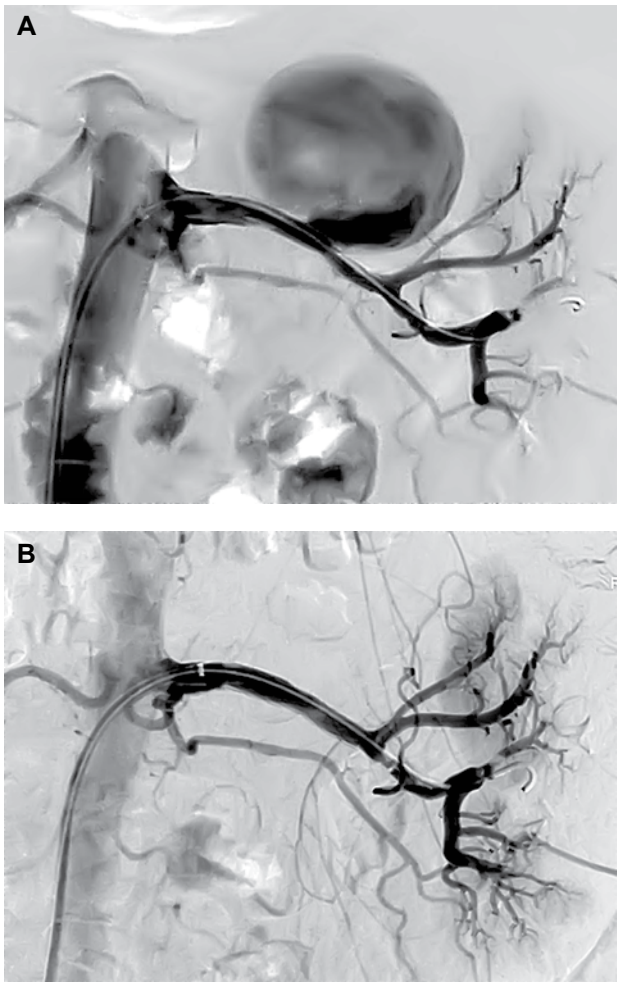

Figure 2. A. An angiographic check confirmed the presence of the 4.6 cm aneurysm. The vessel originating from the aneurysm in the computed tomography scan was not revealed angiographically because of the predilection of flow in the renal artery in the selective study. B. A 5 × 38 mm polytetrafluoroethylene-covered stent was advanced and deployed over the aneurysm neck, successfully excluding the aneurysm.

scan was not revealed angiographically due to the predilection of flow in the renal artery in the selective study. Considering that any exclusion of the aneurysm would not have spared this side branch we decided to deploy a covered stent, sacrificing the branch but excluding the aneurysm rather quickly. A 5 × 38 mm V12 Advanta™ (Atrium Medical Corporation) polytetrafluoroethylene-covered vascular stent was placed over the aneurysm neck and was remodelled slightly with a 6 × 40 mm balloon. Further dilatation of the proximal portion of the stent was performed with a larger 7 × 20 mm balloon, successfully excluding the aneurysm (Figure 2b). An angiographic check of the right renal artery followed and revealed a beaded pattern consistent with FMD. (Figure 3). Following the procedure, urine output fell to 10-20 ml/hr. Urea and creatinine levels rose to 14.6 mmol/l and 355 mmol/l, respectively, from the preoperative values of 6.7 mmol/l and 164 mmol/l. However, renal function improved over the following days, and renal ultrasonography revealed normal echogenicity, and global perfusion of both kidneys. Follow-up investigations indicated normal renal function with an estimated glomerular filtration rate of 78 ml/min. Surveillance CTA three months following intervention showed successful aneurysm exclusion, with blood flow preserved in the left renal artery. Nevertheless, the left kidney now has small areas of infarction (Figure 4) and the patient remains hypertensive on oral antihypertensive therapy.

Discussion

Fibromuscular dysplasia (FMD) is a non-inflammatory, non-atherosclerotic vascular disease of unknown aetiology that affects mainly medium and large vessels, most commonly the renal and internal carotid arteries of young female adults.[4] Approximately 28% of patients have involvement of more than one vascular territory; renal arteries are involved in up to 75% of cases and in half of them involvement is bilateral.[6] In patients affected by FMD, renal artery aneurysms (RAAs) may occur with a prevalence as high as 10%, compared with less than 1% in the non-FMD patients.[7]

Figure 3. Further angiographic examination of the right renal artery also revealed a beaded appearance, consistent with fibromuscular dysplasia.

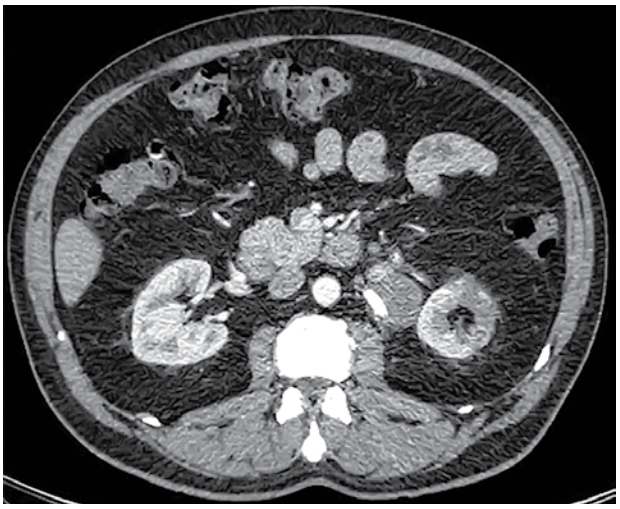

Figure 4. Follow-up computed tomography angiography three months after intervention indicates successful aneurysm exclusion, with blood flow preserved in the left renal artery. The left kidney is smaller and has areas of infarction.

Aneurysms or pseudoaneurysms associated with FMD may occur in any arterial segment involved with FMD, but in the case of the renal artery they are more likely to appear in the middle or distal parts of the main renal artery and the proximal segmental branches. More than 80% are extrarenal, originating at the bifurcation of the main renal artery.[4] RAAs are the fourth most common visceral artery aneurysm after splenic, hepatic, and superior mesenteric artery aneurysms[8] and need to be treated if the diameter exceeds 2 cm because of the higher risk of rupture.

There are several reported cases of ruptured RAAs, and most of these were treated by nephrectomy because of haemodynamic compromise.[9-11]

Endovascular treatment has been reported in two other cases of ruptured RAAs in the literature. Fraser et al[12] report a case of a ruptured RAA in a previously well 45-year-old woman who presented with abdominal pain and syncope. The authors reported that the patient was transferred to the interventional radiology suite and endovascular stenting followed, but gave no further details. Fiesseler et al[13] reported a case of a 52-year-old man with a ruptured right RAA where successful coil embolisation of the proximal right renal artery was performed, but again in this case no follow-up information was included.

In the case reported here, a covered stent was considered the appropriate choice for the treatment of a ruptured RAA. The patient was haemodynamically unstable and the aneurysm had a rather wide neck. The alternative of coil embolisation was considered to be risky, given the possibility of coil migration, and also time consuming in an acute setting. Some authors reported the use of a non-covered stent in order to preserve the position of the coils. Manninen et al[14] performed stent-assisted embolisation of wide-necked renal artery bifurcation aneurysms by placing a dedicated neurointerventional nitinol stent in the aneurysm neck, followed by deployment of detachable coils. This technique is difficult to apply in an emergency setting, whereas with the deployment of a covered stent immediate haemodynamic stability may be achieved. We were obliged to sacrifice a branch of the renal artery, at the cost of infarction of a portion of the kidney in the follow-up scan, but even if coil embolisation had been performed this branch would probably have been sacrificed. In addition, by avoiding nephrectomy, we managed to preserve at least part of the left kidney parenchyma. This is particularly important given the disease in the contralateral renal artery, which may also have been the cause of the patient's residual hypertensive status.

This patient presented with FMD in combination with a ruptured aneurysm. He was haemodynamically unstable and emergency treatment was necessary. The use of a covered stent is a reliable, rapid and effective method in an emergency setting. Although some nephron loss occurred, the overall result was

satisfactory, indicating that endovascular treatment is the first line approach in such cases.

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