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SHORT REPORT

Rupture of a Median Sacral Artery Aneurysm in a Patient with Vascular Ehlers Danlos Syndrome

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Introduction: Vascular Ehlers Danlos syndrome is a rare connective tissue disease that is associated with various arterial complications.
Report: A 25 year old man with vascular Ehlers Danlos syndrome presented with acute lower back pain as a result of a ruptured aneurysm of the median sacral artery (MSA). Prior medical history included several vascular events resulting in a right iliac occlusion. The unusual location of aneurysmal disease of the MSA might be explained by extensive collateral flow recruitment due to this occlusion.
Conclusion: Previous vascular events inducing collateral recruitment might justify a more frequent follow up in patients with connective tissue disorders.

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INTRODUCTION

Vascular Ehlers Danlos syndrome (EDS), formerly known as EDS type IV, is an uncommon subtype of EDS. The disorder is caused by a mutation in the *COL3A1* gene, responsible for synthesis of type III procollagen. Patients affected with vascular EDS are at high risk of vascular events such as dissections, post-dissection aneurysms or arterial rupture, and affected sites may include the aorta or the visceral and peripheral arteries. Management consists of aggressive blood pressure reduction and treatment of acute complications. There are no official guidelines available for vascular follow up.

A case of a ruptured aneurysm of the median sacral artery (MSA) is reported in a patient with vascular EDS.

CASE REPORT

A 25 year old man, diagnosed with vascular EDS at the age of ten following analysis for a possible coagulation disorder, presented at the emergency department with lower back pain. Prior medical history included spontaneous haemothorax, a pyloromyotomy and spontaneous splenic haemorrhage. At the age of 20 he had endovascular treatment for right iliac dissection and rupture, and several months later a right common iliac artery aneurysm was treated with an aorto-unifemoral bypass. Occlusion of the aorto-unifemoral bypass eventually led to an occluded right iliac axis. A ruptured left external iliac aneurysm was treated by coiling of the internal and stenting of the external iliac artery.

At presentation he was haemodynamically stable and laboratory results showed a haemoglobin of 76 g/dL, leucocytes of 19.46 $\times$ 10^9/L, a CRP of 16.3 mg/L and a CK of 327 U/L. Computed tomography angiography (CTA) showed contained rupture of a median sacral artery aneurysm (Fig. 1). The median sacral artery had hypertrophied to a diameter of 5 mm following occlusion of the right common iliac artery. Although open reconstruction remains the treatment modality of choice for vascular pathology in patients with connective tissue disease, given the extensive previous abdominal vascular surgery, an endovascular approach was chosen. Using left brachial access, outflow vessels were occluded using coil embolisation (Boston Scientific Interlock™), following which the aneurysm was filled with fibred coils (Cook MReye®). The origin of the median sacral artery was occluded using a vascular plug (Amplatzer™ Vascular Plug 4, device diameter 7 mm). This was considered to be a better option than coil embolisation since there was only a short normal proximal segment of median sacral artery. Eight hours after the procedure, the patient became haemodynamically unstable due to retroperitoneal re-bleeding. There was persistent proximal filling of the aneurysm and a tapered iliac stent (Medtronic© Tapered 16/13/92 iliac extension) was placed in the distal abdominal aorta, landing in the common iliac artery to

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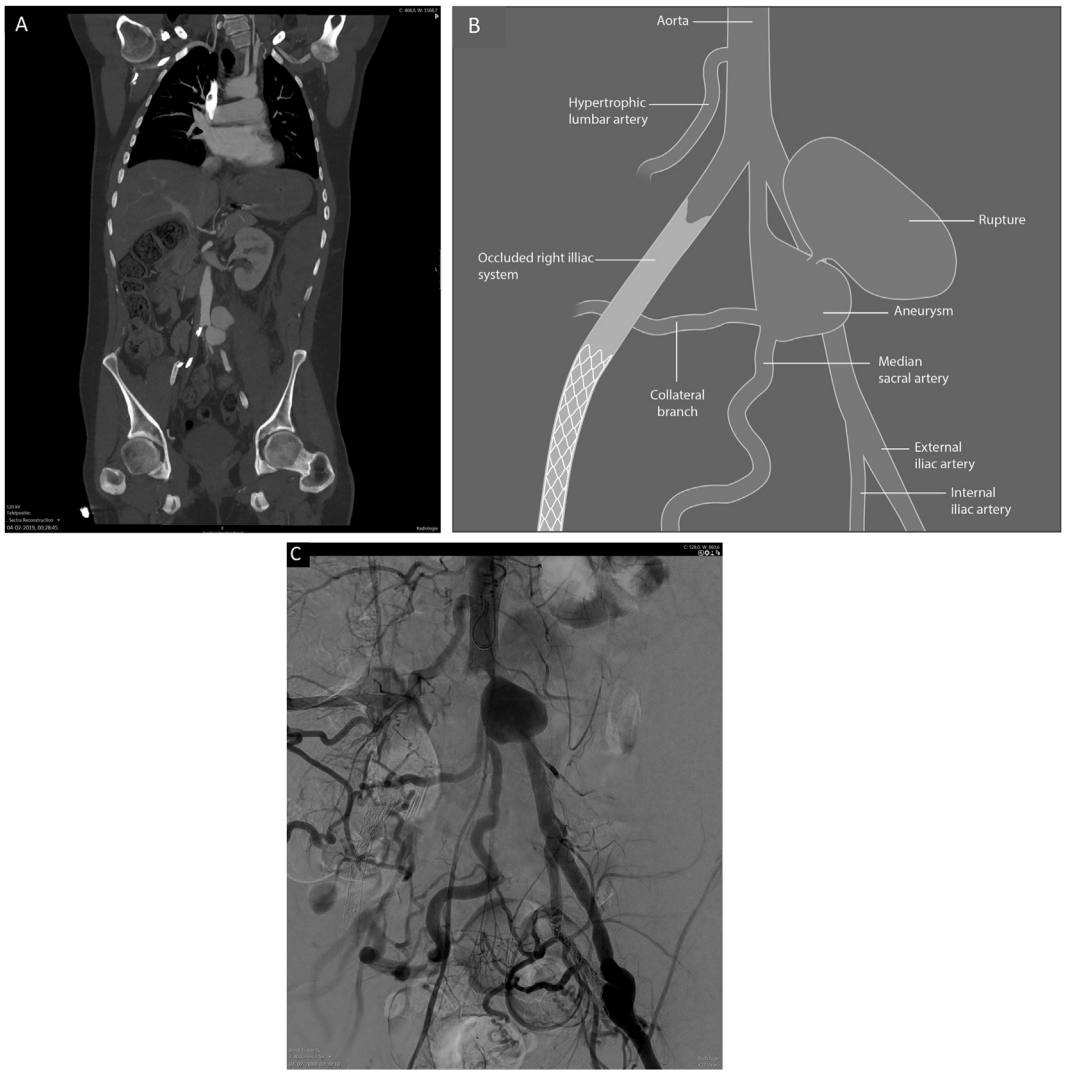

Figure 1. (A) Abdominal computed tomography angiography coronal view demonstrating the aneurysm arising from the median sacral artery. (B) Schematic overview of the ruptured aneurysm (the left internal iliac artery is patent despite previous coiling). (C) Digital subtraction angiography demonstrating the aneurysm.

definitively occlude the origin of the median sacral artery (Fig. 2). The patient rapidly recovered and was discharged after 7 days, without further complications. During follow up, right sided short distance claudication (100 meters) was noted, which will be managed conservatively. Surveillance will be performed by CT angiography and duplex ultrasound within six months and one year after these events.

DISCUSSION

A case of a ruptured median sacral artery aneurysm is reported. This very unusual location might be explained by assuming that increased flow due to collateralisation led to significant changes in flow haemodynamics. A *COL3A1* gene defect leads to diminished arterial tensile strength due to a decreased amount of collagen III. This might have led to rapid aneurysmal growth and rupture.[1] A few case reports describe ruptured aneurysms of the *lateral* sacral artery in patients with a renal transplant.[2,3] It is hypothesised that high flow pelvic collaterals formed due to a relative steal by the renal graft anastomosed to the external iliac artery, subsequently leading to aneurysmal degeneration. Aneurysms are also seen in the pancreaticoduodenal and gastroduodenal artery, secondary to coeliac artery (CA) occlusion or stenosis.[4] As a result of CA occlusive disease, collaterals arising from visceral collaterals, being subject to high flow, can cause dilatation in patients without underlying connective tissue disorders. These findings support the authors' hypothesis on the aetiology of the aneurysm in the present patient. In this case rupture occurred three years following iliac occlusion. A follow up CT angiogram one year before rupture showed a normal median sacral artery. This means the aneurysm developed rapidly, which can be attributed to the underlying connective tissue disorder.

The case raises questions about vascular follow up in vascular EDS and other connective tissue disorders. There are currently no widely accepted guidelines on surveillance in vascular EDS patients. Oderich et al.[5] state that there is an advantage in regular imaging studies. The consensus group by Byers et al.[6] recommends that annual surveillance of the vascular tree by doppler ultrasound, CTA or MRA is

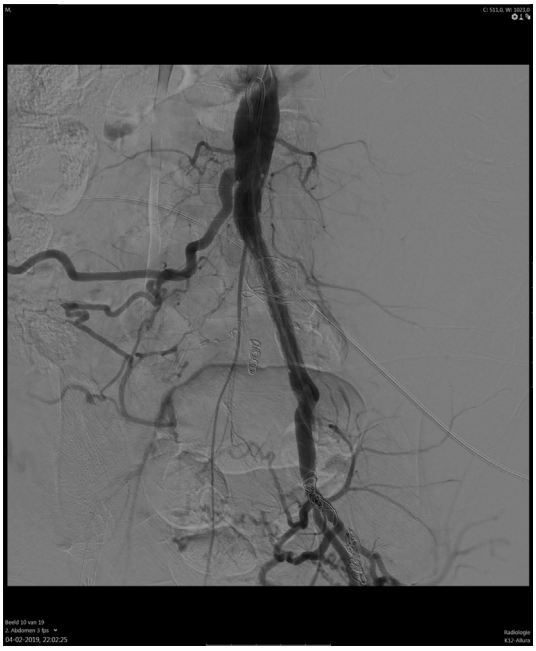

Figure 2. Digital subtraction angiography demonstrating the final result, after placing a stent in the distal aorta landing in the left common iliac artery.

recommended. It must be noted that negative diagnostic imaging does not exclude the risk of non-aneurysmal arterial complications such as dissection. When it is assumed that collateralisation can cause aneurysm formation in patients with connective tissue disorders, aneurysms might develop rapidly, and could be missed by regular follow up. A more frequent follow up in the first years after the vascular event causing collateralisation can therefore be justified. Since it is obvious that elective arterial surgery will be better tolerated in most patients with connective tissue disorders this approach might prevent mortality and morbidity.

CONCLUSION

This case demonstrates a median sacral artery aneurysm in a patient with vascular EDS. The relative rarity of the aneurysm makes it difficult to determine the exact pathogenesis, natural history and optimal management. Increased flow due to collateralisation might cause rapid aneurysmal degeneration. More frequent follow up with non-invasive diagnostic modalities might be justified.

CONFLICTS OF INTEREST

None.

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