CASE REPORT

Left gastric artery aneurysm in Marfan syndrome: a unique case

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Accepted 9 June 2015

SUMMARY

A 78-year-old man presented with abdominal pain, high International Normalised Ratio and rapidly falling haemoglobin. He had a background of Marfan syndrome and was on warfarin following mechanical aortic valve replacement. Abdominal CT imaging showed haemoperitoneum with a leaking aneurysm of the left gastric artery measuring 10 mm in diameter. The decision whether to reverse his anticoagulation provided a difficult clinical scenario given the risk of thrombosis associated with the mechanical aortic valve. The patient went on to have a successful percutaneous embolisation of the aneurysm using a metallic coil and Onyx. Left gastric artery aneurysms are rare and have a reported mortality of up to 70%. This is the first case of gastric artery aneurysm described in a patient with Marfan syndrome.

BACKGROUND

Visceral artery aneurysms (VAA) are rare and have a documented incidence of 0.01–0.2%.¹ Gastric artery aneurysms comprise only 4% of the total number of VAA's and typically present following rupture, which can be fatal in up to 70% of cases.² The majority of gastric artery aneurysms are intramural, classically causing massive gastrointestinal haemorrhage if rupture occurs. The remainder (30%) are extravisceral aneurysms that tend to cause haemoperitoneum when they rupture.²

Marfan syndrome (MFS) is an autosomal dominant connective tissue disorder causing vascular, skeletal and ocular complications. MFS has been linked with cystic medial necrosis affecting large arteries, typically leading to aortic aneurysms.³ However, aneurysms affecting many different arteries have been described in MFS. One study showed that MFS was present in 3% of cases of visceral artery aneurysms.⁴ Despite this, we can find no documented cases of gastric artery aneurysm in Marfan syndrome described in the literature. We therefore believe that this is a unique case and one that posed a number of difficult challenges in diagnosis and management.

CASE PRESENTATION

A 78-year-old man presented with a 3-week history of general malaise. Over this time period he had seen his primary care physician due to feeling increasingly short of breath, for which he was treated with oral antibiotics for a lower respiratory tract infection. He also reported mild generalised abdominal discomfort. He denied any bleeding

and, in particular, denied any haematemesis or melaena.

On the day before presenting to hospital, blood tests showed a high International Normalised Ratio (INR) >10, for which the patient was treated in the community with oral vitamin K. The following day, he was referred directly to the Medical Assessment Unit by his primary care physician, who was concerned that he was looking unwell.

The patient's medical history was notable for Marfan syndrome complicated by aortic root aneurysm for which he received an aortic root replacement and mechanical aortic valve replacement. Several other family members were also affected by Marfan syndrome. He also had a history of atrial fibrillation, chronic kidney disease stage II, total hip and knee replacements and bilateral thoracotomies for bullous emphysema.

The patient's medications, apart from warfarin, included lisinopril, pravastatin, finasteride and co-codamol.

He was an ex-smoker with occasional alcohol intake and was functionally independent.

On examination, vital signs showed a heart rate of 80 bpm, blood pressure 115/62 mm Hg, respiratory rate 20, oxygen saturations 96% on air and temperature of 37.4°C. Cardiovascular examination was remarkable for a prosthetic second heart sound with no added murmurs, and respiratory examination was normal. Abdominal examination revealed a soft, diffusely tender abdomen with no evidence of peritonism.

INVESTIGATIONS

Notable admission blood tests showed: haemoglobin 87 g/L (111 g/L the day before) with a normal mean corpuscular volume (88.1 fL), white cell count 23.9×10^9 , INR 2.0 and an acute kidney injury with estimated-glomerular filtration rate of 36 mL/min/1.73 m². Liver function tests and serum lipase were normal.

Following review in the Medical Assessment Unit, an urgent contrast-enhanced abdominal CT scan was requested. This revealed an aneurysm of the left gastric artery measuring 10 mm in diameter (figure 1). There was also evidence of haemoperitoneum and haematoma of the lesser sac (figure 2). Repeat blood tests at this time showed a fall in haemoglobin to 69 g/L and INR of 1.7.

TREATMENT

After discussion with the local haematology department, the decision was made for further warfarin reversal with vitamin K, given the

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To cite: Powell RO, Babu SB, Bommayya G. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-208342

Figure 1 Abdominal CT showing focal aneurysm and haematoma in upper abdomen.

presence of life-threatening bleeding, despite the risk of thrombosis associated with the patient's mechanical aortic valve. He required intravenous fluid resuscitation and blood transfusion with 2 units of packed red blood cells due to hypotension.

At this point, it was decided that it would be in the patient's best interest to transfer urgently to the local interventional radiology department for percutaneous embolisation. A catheter was fed into the right common femoral artery and advanced through the coeliac axis to the distal left gastric artery where the aneurysm was identified (figure 3). Contrast was demonstrated to be leaking at this site (figure 4). A metallic coil was inserted into the aneurysm. However, contrast was still shown to be flowing to the aneurysm, possibly due to the fact that the patient was anticoagulated. Further embolisation was attempted with administration of Onyx (liquid embolisation agent) into the aneurysm. This led to successful elimination of blood flow to the aneurysm, however, a small paradoxical embolisation of the left hepatic artery was noted, which was not flow limiting and therefore of no clinical significance.

Figure 2 Abdominal CT showing haemoperitoneum.

Figure 3 Angiogram showing leaking left gastric aneurysm.

OUTCOME AND FOLLOW-UP

Following the embolisation procedure, the patient was transferred to the ward for further management. His blood pressure stabilised following transfusion of 2 units of packed red blood cells. Repeat blood tests showed a rise in haemoglobin to 98 g/L.

The following day, the decision was made to restart anticoagulation, initially with intravenous unfractionated heparin to ensure rapid reversibility in case of further bleeding. The patient was subsequently restarted on warfarin. He continued to improve and was discharged home 2 weeks later.

DISCUSSION

This case highlights a difficult clinical scenario with life-threatening intra-abdominal bleeding occurring in a patient on warfarin for mechanical aortic valve replacement. Warfarin reversal increases the likelihood of valve thrombosis and

Figure 4 Angiogram after embolisation shows complete elimination of flow to aneurysm.

thromboembolic events. Despite this, high-dose vitamin K with or without fresh frozen plasma is the recommended treatment in patients with mechanical valve replacement and warfarin-induced major bleeding.⁵

Minimally invasive endovascular embolisation of VAA's is increasingly used and has been well documented.⁶ In particular, glue embolisation of a left gastric artery aneurysm has previously been documented with good results.⁷ One series showed a 98% initial success rate of minimally invasive intervention for visceral artery aneurysms. The rate of rebleeding requiring further intervention within 30 days in this series was only 3%.⁸

We hope that this case highlights a number of interesting clinical issues. Aneurysms of the left gastric artery are rare and have a high reported mortality when they rupture. Without timely intervention, it is likely that the outcome in this case would have been poor.

Learning points

- ▶ Aneurysms of the left gastric artery are rare and can be life-threatening if they rupture.
- ▶ Visceral artery aneurysms should be considered in patients with Marfan syndrome or other connective tissue disorders who present with abdominal pain.
- ▶ Percutaneous embolisation is a recognised and effective treatment option.

Acknowledgements Thanks go to Dr Mike Williams who was the reporting Radiologist on this case.

Contributors ROP wrote the primary manuscript. SBB provided the images. SBB and GB reviewed and corrected the paper before submission.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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