A Challenging Case of Fibromuscular Dysplasia in a Transgender Patient: Is There a Hormonal Link?

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Introduction: Fibromuscular dysplasia (FMD) and superior mesenteric artery (SMA) aneurysms are rare vascular conditions. An unusual combination of both diseases is reported.

Case report: A 54 year old woman presented with symptomatic SMA aneurysm. A diagnosis of FMD was made on the basis of computed tomography angiography (CTA). The patient had undergone gender reassignment surgery 10 years previously and continued to use both topical and oral hormonal therapy. The patient received open anatomical bypass through a retroperitoneal approach using great saphenous vein.

Conclusion: Superior mesenteric artery aneurysms are rare and a diagnosis of FMD should be considered as part of the diagnosis process. Anatomical bypass should be considered carefully in relation to a patient’s fitness as well as anatomical suitability.

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INTRODUCTION

Fibromuscular dysplasia (FMD) is a rare disease defined as thickening of the arterial media by fibrosis and muscular hyperplasia, causing multifocal arterial stenosis. It most commonly affects the renal artery (80%) and then the internal carotid artery, but can be found in any artery in the body. FMD predominantly affects females. The aetiology of FMD is unknown; however, associated factors include tobacco, oestrogen, and genetic factors.

Superior mesenteric artery (SMA) aneurysms are rare, with a prevalence of less than 7% of all visceral artery aneurysms. They usually present as incidental findings on cross sectional imaging, but may present acutely as a rupture, or as bowel ischaemia. Atherosclerosis is the usual aetiology; however, FMD may present as an SMA aneurysm.

In this case report, the challenges of an acute presentation of a SMA aneurysm secondary to FMD are presented, and the potential association between FMD and gender reassignment is explored.

REPORT

A 54 year old woman was under general surgical outpatient investigation for an 8 week history of loose stools, increasingly severe abdominal pain after eating, and 19 kg weight loss. Initially, examination confirmed a soft non-tender abdomen but over the weeks her symptoms became progressively more constant and she developed diffuse abdominal tenderness. She had a background of smoking, and had had gender reassignment surgery 10 years previously, and had been on oral hormonal therapy which had stopped. Currently, she was on topical oestrogen gel 0.06% once daily as well as oral anti-androgen finasteride 2.5 mg once daily. She was admitted for investigation at her local hospital. A portal phase contrast computed tomography (CT) scan of abdomen and pelvis reported a probable acutely thrombosed SMA aneurysm. She was transferred to the local vascular surgery unit and formally anticoagulated with unfractionated heparin, she underwent a second arterial-venous phase CT abdomen/pelvis and urgent endoscopy. This demonstrated multiple aneurysms of the visceral arteries. These included a 12 mm aneurysm of the coeliac artery, 9 mm aneurysm of the splenic artery, small aneurysms of the distal renal arteries, and a 16 mm aneurysm of the SMA. The SMA aneurysm was located 1 cm from its origin and associated with partial thrombus (with no acute features) before occluding. The anatomy was complicated by an aberrant right hepatic artery arising from the SMA origin immediately proximal to the aneurysm and was the dominant supply to the liver and the gastroduodenal artery (Fig. 1). The appearance of both renal arteries demonstrated a beaded appearance with sparing of the ostia diagnostic of FMD (Fig. 1). In addition, there were non-flow limiting dissections of the common, internal, and external iliac arteries. The appearances were strongly suggestive of fibromuscular dysplasia. A CT angiogram of the
carotid and cerebral arteries showed a 6 mm aneurysm of the mid right internal carotid artery with the typical of the appearances of FMD (Fig. 1). Access to the origin of the SMA anterior to the left kidney was achieved by reflection of the spleen. The distal SMA was controlled beyond aneurysm via the infra-coolic compartment. A length of great saphenous vein was harvested and reversed. An SMA aneurysm bypass was performed anatomically, using the inlay technique, from the origin of the SMA (retroperitoneal) to the distal SMA beyond the aneurysm (infra-coolic compartment). The graft was tunneled anatomically through the root of mesentery (Figs. 2 and 3).

The patient made a good recovery and was discharged 11 days post operatively. A CTA demonstrated technical success (Fig. 3). Routine post-operative follow up included outpatient review at 6 weeks and duplex ultrasound of the graft at 12 weeks. No further graft surveillance was planned. FMD surveillance with an MRA of the carotid, circle of Willis, and visceral segment of the aorta on alternate years, is planned. A discussion was undertaken with the patient regarding the potential association between hormonal therapy and FMD. The patient declined to discontinue or reduce hormone supplements.

DISCUSSION

FMD is currently defined as an “idiopathic, segmental, non-inflammatory and non-atherosclerotic disease of the musculature of arterial walls, leading to stenosis of small and medium-sized arteries,” which may be familial. Kadian-Dodov et al. reported that 41.7% of patients had a diagnosis of an aneurysm and/or dissection. Registry data have shown that the extracranial carotid and vertebral arteries were nearly as frequently involved as the renal arteries. Although of unknown aetiology, smoking and genetic factors have been linked with the disease, as well as an association with oestrogen exposure and increased numbers of progesterone receptors. In this case report, although the patient was XY, it is suggested that there may be a link between gender re-assignment hormone therapy and the development of FMD. A literature search revealed no similar reported cases, but the number of gender re-assignments is increasing and the levels of oestrogens in this population are typically many times the physiological normal in the XX population. This case report adds to the weight of evidence of a hormonal component in the aetiology of FMD, however more research is needed in this area.

The European Society of Vascular Surgery recently published guidelines for the management of diseases of the mesenteric arteries. A class one recommendation was the urgent repair in patients with symptomatic true aneurysms of the mesenteric arteries, irrespective of size or location. Treatment for SMA aneurysms is well described with both endovascular and open repair options. Suitability for
endovascular repair relies on the anatomy of the lesion and other considerations such as age and comorbidities, and the longevity of any repair should be considered. In this case, the patient was treated by an anatomical repair using great saphenous vein. The advantages were perceived to be preservation of an aberrant right hepatic artery, a shorter anatomical bypass, improved long-term patency, and reduced infection risk. The approach to this repair could have been from the front through the infra-colic compartment or using a medial visceral rotation to approach the retroperitoneal aorta at the level of the visceral arteries, the advantage of the latter being access to the origin of the SMA with the option of debranching off the aorta if needed. The young age and good general health of the patient made the additional risks and morbidity of this procedure acceptable.

CONCLUSION
This case report cannot confirm the association between the oestrogen hormonal treatment and FMD. However, the importance of this potential association is only likely to grow, and further research is needed.

SMA aneurysms are a rare and challenging problem. All treatment options should be considered, including open repair.

CONFLICTS OF INTEREST
None.

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