CASE REPORT

Spontaneous hepatic artery dissection—a rare presentation of fibromuscular dysplasia

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Abstract

Fibromuscular dysplasia (FMD) is a rare condition that causes structural compromise of the blood vessel presenting either as an incidental radiological finding, dissection or stenosis usually of the renal or craniocervical arteries. Seldom, patients present with spontaneous dissection in visceral arteries and there are few reports of hepatic involvement. This report outlines the case of a 43-year-old female who presented with severe right upper quadrant pain with a subsequent diagnosis of FMD manifesting as spontaneous hepatic artery dissection. The patient was treated with conservative antiplatelet therapy and regular radiographic follow-up, decided by the treating team as no clear guidelines exist for management of this particular presentation of FMD. Surgical management is not currently recommended to this patient due to the risk of further dissection, but may be considered if there is severe haemodynamic compromise or refractory pain.

INTRODUCTION

Fibromuscular dysplasia (FMD) is a rare condition that primarily affect the renal and cervicocephalic arteries. To the best of our knowledge, only three FMD cases have been reported to present with a spontaneous hepatic artery (HA) dissection, the last in 1994 [1–3]. There have been 27 cases of isolated HA dissection of any cause, most result in surgically managed aneurysms or are discovered incidentally on autopsy [4, 5]. Only one other reported FMD-related HA dissection was successful with medical treatment as our patient was [3].

INTRODUCTION

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CASE REPORT

We present the case of a 43-year-old Caucasian female with newly diagnosed FMD presenting as spontaneous isolated HA dissection. Her history is significant for minimal trauma dissections, an iatrogenic left main stem coronary artery (LMA) dissection during a coronary angiogram for investigation of non-ST elevation myocardial infarction in 2007; this was treated emergently with dual vessel arterial bypass grafting.

Then in 2001, an internal carotid artery dissection following intraoperative arterial line placement during a transphenoidal surgery for Cushing’s syndrome secondary to adrenocorticotropic hormone (ACTH)-secretory microadenoma.

She was diagnosed with medically managed primary hypertension at 19 and also has a history of recurrent transient ischaemic attacks (TIAs) (Fig. 1). She has an extensive medical and surgical history that is largely non-contributory (Table 1).

She was admitted for investigation and management of a TIA presenting as expressive dysphasia and left hemiparesis which developed following cardiac stress testing. While inpatient, she developed sudden severe right upper quadrant (RUQ) abdominal pain associated with dizziness and hyperventilation.

Following her initial normal liver function test (LFT), a subsequent LFT demonstrated mixed hepatocellular derangement which improved over 3 days (Table 2).
Figure 1: Fusiform dilatation of the internal carotid artery suggestive of chronic dissection (arrow).

Table 1: Patient demographic and complete previous medical and surgical history

| Patient demographics | Gender: female  | BMI: 33.1 |
|----------------------|-----------------|-----------|
| Age: 44              |                 |           |
| Medical condition    | Year            | Surgical conditions/procedures |
| Hypertension         | 1991            | Transsphenoidal resection of microadenoma |
| Tendonitis           | 2000            | Renal calculus |
| Cushing’s disease (ACTH microadenoma) | 2000 | Cholecystitis/cholecystectomy |
| Hypercholesterolaemia | 2001        | Umbilical hernia |
| Non-alcoholic fatty liver disease | 2001 |
| Depression/anxiety | 2001            |
| Bronchial asthma     | 2001            |
| Gastro-oesophageal reflux disease | 2002 |
| Irritable bowel syndrome | 2002 |
| Primary osteoarthritis | 2002 |
| Obstructive sleep apnoea | 2003 |
| Ischaemic heart disease | 2006 |

Infectious cause

| Epstein barr virus (EBV) serology | IgG reactive |
| Cytomegalovirus (CMV) serology   | IgG reactive |
| Human immunodeficiency virus (HIV) serology | IgM non-reactive |
| Hepatitis A viral serology       | Non-reactive |
| Hepatitis B viral serology       | Non-reactive |
| Hepatitis C viral serology       | Non-reactive |
| Syphilis (EIA) total antibody     | Non-reactive |

Toxic cause

| Paracetamol (acetominophen) level (mg/L) | <10 |
| Autoimmune cause                       |     |
| Antinuclear antibody (ANA)             | 1:160 speckled |

Continued
After numerous surgical reviews, an unremarkable abdominal ultrasound, plain abdominal computed tomography (CT), stable haemoglobin (Hb), negative viral hepatic serology and effective pain relief from simple and opiate analgesia; it was presumed that the liver injury was related to inpatient substitution of rosuvastatin for atorvastatin due to restricted supply. With substantial improvement in LFTs, she was discharged. She represented 6 days later complaining of severe intermittent RUQ pain with a worsened mixed hepatocellular LFT derangement, various medical causes were excluded (Table 2). Magnetic resonance cholangiopancreatography (MRCP) found abnormal wall thickening suspicious of dissection of coeliac trunk and HA (Fig. 2). Mild stenosis was demonstrated on abdominal doppler ultrasonography and prompted a confirmatory abdominal CT-Angiogram. Dilatation of the common HA and presence of an intimal flap with associated fat stranding was consistent with dissection (Fig. 3). No dissection was

![Figure 2: MRCP demonstrating abnormal wall thickening (arrow).](https://academic.oup.com/omcr/article-abstract/2016/11/omw083/2557043/2071042)
found in the coeliac trunk. The biliary tree was normal and no thrombosis or haemorrhage was discovered (Figs 4 and 5).

Multiple specialty teams agreed to conservative management consisting of dual-antiplatelet therapy, ambulatory blood pressure monitoring, six monthly abdominal imaging and lifestyle modification regarding high impact activity. Consensus was that anticoagulation was not indicated.

The patient is currently alive but has represented repeatedly with similar symptoms that was not deemed for surgery. A follow-up abdominal US four months after the index event demonstrated progression of HA flow from 89 to 270 cm/s with a pseudoaneurysm of 6 mm, repeat CT has confirmed the dissection is still patent.

**DISCUSSION**

Regardless of aetiology, HA dissections present similarly with acute abdominal pain affecting the epigastrium, RUQ and back [6]. Most are discovered incidentally with various associated symptoms. Investigations commonly demonstrate deranged LFTs; however, isolated cases report normal biochemistry despite disease acuity [7].

The patient in this case had deranged LFTs without clear medical cause (Table 2). No intrahepatic aneurysm, haemorrhage, thrombosis or fistulization was noted on any subsequent imaging, excluding haemobilia. An intimal flap and aneurysm was ultimately located in close proximity to the bifurcation of the common HA causing relative ischaemia and correlates with her LFT pattern.

FMD can be diagnosed based on histologic or radiographic criteria. Histologic diagnosis is uncommon due to significant complications in specimen acquisition and advanced radiographic techniques. Alternating areas of stenosis and aneurysmal dilatation is atypical of pathology such as atherosclerosis and vasculitis, and serology was negative. Segmental arterial medialysis is an important differential diagnosis but is radiologically difficult to differentiate from FMD. Multiple specialist radiologists concluded the diagnosis of FMD especially given her history of cerebrovascular dissection.
Approximately two-thirds of subjects who underwent imaging in the US FMD registry had more than one vascular bed affected by FMD [8]. Our patient has known carotid and LMA dissections in addition to the common HA and these dissections could be at least in part due to underlying FMD; however, this possibility was not explored at the time. Hormonal influences have been suggested to be a cause of FMD given the propensity for female sex; however, no relationship between FMD and ACTH levels have been established.

Other than dissection of visceral arteries she is at higher risk of further cerebrovascular and coronary events which reflects findings in the US FMD registry (13.4% TIA, 9.8% stroke and 6.5% coronary events) [8]. Given the patient’s existing ischaemic heart disease, her risk of coronary event is likely to be higher than most [8].

Current FMD registries demonstrate empirical treatment with antiplatelet therapy for stable cases [8], and reports of visceral dissection unrelated to FMD advocate either antiplatelet or anticoagulant therapy, without consensus on the most appropriate first line therapy [7, 9]. No trials demonstrate the risks and benefits of either treatment in visceral artery dissections, and randomized trials analysing dissections in other arteries, particularly cervical, have not found one to be more effective in improving overall outcomes [10].

As treatment with antiplatelet therapy is used in both non-FMD and FMD dissections regardless of location, this was appropriate as no specific guidelines for FMD-related visceral artery dissections exist. Surgery is indicated if the dissection becomes life-threatening or medically refractory pain is present. The patient and surgical team made a joint decision to forego surgery as its risk outweighed the risk of dissection.

There is great need for reporting of visceral artery dissections, especially those related to FMD to appropriately guide clinical decisions. Albeit uncommon, dissection should be considered as a differential in sudden onset epigastric pain without obvious cause. Management should be guided by severity and vigilance is imperative to prevent mortality from vessel rupture; an indication for surgical management.

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CONFLICTS OF INTEREST STATEMENT

None declared.

ETHICS APPROVAL

Ethics Exemption applied for and granted by the National Health and Medical Research Council (NHMRC) at the Royal Brisbane and Women’s Hospital. File Reference number: HREC/16/QRBW/80. Contact: Level 7, Block 7, Butterfield St, Herston, Queensland, Australia, 4029. Ph: +61 3646 5490. Facsimile: +61 3646 5849. E-mail: RBWH-Ethics@health.qld.gov.au

CONSENT

Consent obtained and included.

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