Hepatic and mesenteric fibromuscular dysplasia: an uncommon entity

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Abstract

Dissection of the common hepatic artery is a rare cause of acute abdomen. In this case report, we discuss a 58-year-old woman who presented with severe epigastric and back pain, for which she had an abdominal computed tomography (CT) scan. On CT images, a dissection of the common hepatic artery, extending to its right and left dividing branches and a diffusely pathological aspect of the hepatic and digestive arterial network was noted. A diagnosis of extensive fibromuscular dysplasia (FMD) was suggested. FMD is a non-atherosclerotic, non-inflammatory arterial disease that most commonly involves the renal and extracranial carotid arteries, but almost all arterial beds may be affected. There is a high prevalence of aneurysm and dissection among patients with FMD.

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Case summary

A 58-year-old woman presented to our emergency department with complaints of increasing worsening epigastric and back pain for two days. She also had nausea and vomiting. She denied fever, chills, chest pain, dyspnea, cough, rectal bleeding and mictaligia. Review of systems was otherwise negative. The patient suffered from migraines, arterial hypertension and had history of carotid aneurysm.

Physical examination of the patient was performed in the emergency department and revealed an otherwise well-appearing patient. Abdominal examination revealed a supple and depressible abdomen with a diffuse tenderness and an epigastric soreness. Peristalsis was normal. There was no re-bound, pulsatile, or palpable mass. Physical examination was otherwise unremarkable.

The patient was given an intravenous opioid painkiller for pain control.

Laboratory results obtained showed neutrophilic hyperleukocytosis, mild hyperlactatemia, increased fibrinogen and slightly disturbed liver enzymes. The rest of the blood biology was ordinary.

Imaging findings and diagnosis

An abdominopelvic computed tomography (CT) with IV contrast performed at the hospital revealed an aneurysmal dila-
Fig. 1 – (B) The “string of beads” appearance of intrahepatic branches. (D) Alternating stenosis and dilatation of the pancreaticoduodenal arteries. (F) Dissection of the common hepatic artery with an intimal flap.
Fig. 2 – (A/B) The “string of beads” appearance of intrahepatic branches. (C) Intrahepatic artery dissection. (D) Alternating stenosis and dilatation of the pancreaticoduodenal arteries. (E) Dissection of the inferior mesenteric artery.
Fig. 3 – Renal arteries caliber irregularities.
tion of the celiac trunk secondary to juxta-ostial stenosis due to the median arcuate ligament. There was also, just downstream from the emergence of the splenic artery, a dissection of the common hepatic artery, extending to its right and left dividing branches (Fig. 1). There was no objective aterial occlusion. There was no extension to the gastroduodenal, splenic and left gastric arteries.

In addition, a diffusely pathological aspect of the hepatic and digestive arterial network was noted: multiple vascular irregularities such as alternating stenotic and dilation segments (Figs. 1–2). This aspect suggested systemic vasculopathy. A similar aspect affected the renal arteries (Fig. 3). The dissection appeared partially thrombosed in the proximal part of the inferior mesenteric artery (Fig. 2).

A diagnosis of extensive fibromuscular dysplasia was suggested, and the therapeutic options include antiplatelet, anticoagulant and antihypertensive therapy.

Discussion

Fibromuscular dysplasia (FMD) is a non-atherosclerotic, non-inflammatory, systemic disease of small- to medium-sized arteries, that may result in arterial beading, stenosis, occlusion, aneurysm, or dissection. FMD mainly affects the renal and extracranial carotid and vertebral arteries, but nearly all vascular beds may be affected, and multivessel involvement is common. The extracranial carotid, renal, and intracranial arteries were the most common sites of aneurysm when dissection most often occurred in the extracranial carotid, vertebral, renal, and coronary arteries [1–4]. Although a variety of genetic, mechanical, environmental and hormonal factors have been proposed, the mechanism underlying the pathogenesis of FMD remains poorly understood [1–6].

Most patients with this condition are women, frequently diagnosed in middle age, and present symptoms, depending on the vessels involved, with hypertension, migraine, or complications as such as RA or cerebrovascular event (transient ischemic attack, dizziness, stroke, subarachnoid hemorrhage, ...). The prevalence of FMD is not precisely known because this disease is often clinically silent or discovered incidentally [2,3,6]. In the US registry, mesenteric ischemia was uncommon manifestation of FMD, reported in only 1.8% of patients and the celiac and mesenteric arteries accounted for 6.8% of all arterial dissections and 22.3% of all arterial aneurysms reported [1].

FMD is currently classified by angiography into two subtypes, multifocal and focal. The presentation of multifocal FMD is a typical string-of-beads pattern and is at least 4 × more frequent than focal FMD [7]. By the way, visceral artery FMD includes the celiac axis, hepatic, splenic arteries, and the superior and inferior mesenteric arteries. Patients with visceral FMD are more likely to have aneurysms or dissections compared to those without visceral FMD [5,7,8]. The clinical manifestations of FMD are variable and depend on the distribution of vascular beds involvement, the type and severity of arterial lesions [1]. Specifically, clinical symptoms of mesenteric FMD include postprandial abdominal pain, weight loss, fatigue, incomplete intestinal obstruction, focal or multiple ulcers of the intestinal mucosa, and abdominal distension.

It is recommended that every patient with FMD undergo one-time cross-sectional imaging from head to pelvis with computed tomographic angiography or magnetic resonance angiography. The factors that distinguish atherosclerosis from FMD include older age, traditional cardiovascular risk factors, and lesions affecting mainly the ostium and proximal parts of the arteries involved [1,2,4,5]. As opposed to vasculitis like giant cell, Takayasu arteritis, polyarteritis nodosa, Behcet’s disease, defined by marked inflammation, FMD is a non-inflammatory process [1].

Computed tomographic angiography, magnetic resonance angiography and duplex ultrasonography is more commonly used to diagnose and monitor the disease [1,3,6,9]. Arteriography is reserved for cases in which there are diagnosis doubts, to measure the pressure gradient across the stenotic lesion or for endovascular treatment [5,6,8,9].

Treatment of FMD depends upon symptoms as well as the nature and location of arterial lesions, but may include both medical (blood pressure control, antiplatelet agents) and interventional therapies (angioplasty, stents, coils, surgery). In the absence of contraindication, antiplatelet therapy is reasonable for patients with FMD to prevent thrombotic and thromboembolic complications. Smoking cessation should be strongly encouraged for all patients with FMD who continue to smoke [1,5,10].

A regular follow-up imaging should be performed at six months from diagnosis in symptomatic patients and it is also recommended once yearly for asymptomatic patients. Follow-up includes clinical assessment, assessment of renal function (for renal artery FMD), and imaging. At this time, there is insufficient data to recommend specific algorithms for modality and frequency of imaging studies in the follow-up of FMD. The timing of follow-up imaging should be customized to each patient’s pattern and severity of disease, including the need for monitoring of aneurysms or dissections or following revascularization, as well as local imaging resources and experience [2,7].

Conclusion

Fibromuscular dysplasia is a non-atherosclerotic, non-inflammatory arterial disease that most commonly involves the renal and extracranial carotid arteries, but almost all arterial beds may be affected. Multifocal stenoses with the ‘string-of-beads’ appearance are observed at angiography in more than 80% of cases. FMD may be clinically silent but many patients present with hypertension, migraine, stroke, transient ischemic attack, subarachnoid hemorrhage, ... There is a high prevalence of aneurysm and dissection among patients with FMD. Treatment of FMD include both medical (blood pressure control, antiplatelet agents) and interventional therapies (angioplasty, stents, coils, surgery). A regular follow-up imaging is recommended.
Patient consent

We confirm that written, informed consent for publication of this case was obtained from the patient. Zoé Ciccarese, David Byl, Vincent Scavée

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