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Clinical Features and Endovascular Management of Iliac Artery Fibromuscular Dysplasia

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Abstract

Purpose—To identify the spectrum of clinical presentation of iliac artery fibromuscular dysplasia (FMD) and to evaluate the outcomes of endovascular management of iliac FMD for claudication.

Methods and materials—All patients in our institution with a diagnosis of FMD between January 1980 and December 2010 were identified. 14 patients were found to have FMD of the iliac arteries. Associated risk factors included hypertension (79%), hyperlipidemia (64%), smoking history (36%), coronary artery disease (21%), diabetes (0 %), and obesity (36%).

Results—Eight (57%) patients were incidentally found to have iliac FMD on imaging. 6 (43%) patients had life style limiting claudication involving one or both extremities. All 6 patients were reported as mild peripheral arterial disease (PAD) based on ankle brachial index (ABI) measurements (0.7 to 0.9). These six patients underwent 10 endovascular procedures for claudication including angioplasty (n=8) and self-expanding stent placement (n=2). Mean symptom free survival was 56.3 months.

Conclusion—Iliac FMD may be found incidentally or may present with disabling claudication that is amenable to endovascular treatment.

Keywords

Fibromuscular dysplasia (FMD); claudication in FMD; Iliac Artery FMD

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Introduction

FMD is a nonatherosclerotic, noninflammatory vascular disease that most frequently affects renal and carotid arteries. However, involvement of nearly every artery has been reported (1–12). The clinical presentation of FMD may vary from an asymptomatic condition to a multisystem disease that mimics necrotizing vasculitis, depending on the arterial segment involved, the degree of stenosis, and the type of fibromuscular dysplasia(2). Medial fibroplasia is the most common histological type of FMD showing the characteristic string of beads on imaging (Figure 1).

FMD involving the iliofemoral arteries is very uncommon with limited case reports in the literature (13–23). Most often, it is found incidentally in asymptomatic patients undergoing arteriograms or alternative imaging studies for other reasons. Patients may present with intermittent claudication, critical limb ischemia, or peripheral microembolism that manifests as pain and cyanosis in the toes. The clinical presentation of iliac artery FMD may mimic other more common arterial processes like atherosclerosis and vasculitis. Non-invasive studies of the lower extremities such as ankle brachial index (ABI) can be misleading as the resting ABI is often interpreted as mild disease. Diagnosis is usually made on the basis of imaging tests including duplex ultrasound, CT Angiography, or catheter-based digital subtraction arteriography.

Most published literature is limited to anecdotal reports and treatment decisions are based on clinical symptoms. Large case series or controlled trials to guide diagnostic and therapeutic alternatives are lacking. Therefore, we conducted a retrospective study of all patients diagnosed with FMD of the iliac arteries at a single center. The purpose of the present study is to identify the spectrum of clinical presentation of iliac artery fibromuscular dysplasia (FMD) and to evaluate the outcomes of endovascular management of iliac FMD for claudication.

Methods and Materials

Institutional review board approval was granted for this study. All patients with a diagnosis of iliac artery FMD who underwent evaluation at our institution between January 1990 and December 2010 were included. Patients were identified using the electronic medical records at our institution. Key words used for the search included Iliac FMD, Iliac artery fibromuscular dysplasia and FMD of the iliac arteries. Baseline and follow-up data were obtained from the electronic medical records through July 15, 2011.

Computed tomography (CT) angiography

A 16 or 64 multi-channel CT scanner (General Electric, WI) was used. Non-ionic iodinated contrast material (Visipaque 320, GE Healthcare, Princeton, NJ) was intravenously injected with dosing based on the patients weight and renal function ranging between 100–150 mL. Axial, multiplanar, and 3-D reconstructed images were used in the interpretation. Stenoses were quantified based on percent of cross sectional luminal narrowing with respect to the normal reference vessel diameter.

Common iliac, external iliac, internal iliac artery treatment

All procedures were performed according to the standard technique after informed consent was obtained. Depending on anatomic considerations, access was obtained from either an ipsilateral or contralateral common femoral approach. Typically, a 6F–8F vascular sheath (Terumo, Somerset, NJ) was placed in the artery over a 0.035-inch guidewire and the patient was systemically heparinized (50 U/kg). A variety of standard angioplasty balloons and stents were used. Two vessels were treated with two self-expanding stents in the external iliac artery- S.M.A.R.T. (Cordis Corporation, Bridgewater, NJ) or Protégé stent (ev3 Endovascular Inc., Plymouth, MN). The length and diameter of the stent used was based on the length of the stenosis and the diameter of the non-diseased artery distal to the stenosis. Pressure gradients were obtained only if there was a question of lesion severity.

After the procedure, all patients were started on anti-platelet therapy using either 81-mg daily oral dose of aspirin or 300-mg initial oral dose of Clopidogrel (Sanofi-Aventis, Bridgewater, NJ), followed by 75 mg daily oral dose for at least 6-weeks.

Follow-up

Follow-up after endovascular intervention was advised at 1 month, and then at 3, 6, and 12 months, and then every 6 months thereafter. At follow up, clinical evaluation was performed and ABIs were obtained at the discretion of the clinician. If indicated, additional follow-up was obtained including CT angiography, or conventional angiography as needed to determine the patency of the treated arteries.

Technical success of the procedure was defined by less than 30% residual stenosis by visual estimate of the completion angiogram. Clinical success of the procedure was defined as freedom from claudication, absence of restenosis, or revascularization of the target artery after index procedure at 12 months from the date of the initial procedure.

Results

In the present study, 14 patients were identified who had FMD involving the iliac arteries. Two patients were diagnosed in the 1990s (1994 and 1996) and twelve were diagnosed between 2000 and 2010. Eight (57%) were diagnosed incidentally during imaging performed for other reasons. Six (43%) patients presented with claudication and these patients underwent a total of 10 iliac artery angioplasty or stent placement procedures. The earliest intervention was performed in 1999 and the latest in 2010. Demographic and clinical characteristics of the patients are outlined in Table 1. The mean age at diagnosis was 56 years (range 38–70; 86% female).

The cardiovascular risk factors included hypertension (n=11, 79%), hyperlipidemia (n=9, 64%), smoking history (n=5, 36%), coronary artery disease (n=3, 21%), cerebrovascular disease (n=3, 21%), and obesity (n=5, 36%). The majority of the patients had FMD in the renal arteries (n=12).

All 14 patients had external iliac artery involvement with FMD while only one patient had internal iliac artery involvement as well and one other patient had common iliac artery

involvement. Of the 6 patients that underwent endovascular treatment, clinical success) with symptom resolution was achieved in 83% (n=5 and one patient (17%) continued to have bilateral leg pain despite two procedures. This patient was eventually diagnosed as having neurogenic leg pain. Two patients required stent placement due to a persistent aortoiliac pressure gradient. Details of endovascular procedures performed and their outcomes are shown in Table 2. The average follow-up period was 63 months (range: 5–140 months) and there were no complications.

Discussion

A histological classification for FMD was introduced in 1971 by Harrison and McCormack based on the layer of the arterial wall which was involved (24). Of the histological subtypes, medial FMD is the most common type seen in 80–90% of patients (1, R24). Since pathological specimens are not usually available, imaging with catheter angiography remains the standard tool for diagnosing FMD although CTA or MRA are being increasingly used for diagnosis. Caution must be exercised so as to not misdiagnose standing waves as FMD. FMD typically presents as beading in the external iliac arteries rather than the superficial femoral arteries, where standing arterial waves are more commonly seen (25).

Literature on FMD involving the iliac arteries is limited to isolated case reports and small case series (Table 3). It is usually asymptomatic but when symptomatic, the spectrum of clinical presentation can range from acute limb ischemia due to dissection, emboli leading to episodic digital ischemia to chronic ischemia with gradual onset of claudication (15)

Several prior reports have described open surgical treatment with resection of the affected arterial segments with or without bypass grafting and concomitant dilatation of the affected segments (13, 15). More recently, treatment of FMD of the iliac artery has been performed using angioplasty and the majority of the reports are small single center case studies (19).

An English language literature review using Pubmed was performed from January 1983 to December 2012 of all studies which reported treatment of iliac artery FMD (Table 3). The search terms included “iliac artery FMD”, “iliac artery FMD treatment”, and “endovascular interventions for iliac FMD”. Thirteen studies were identified in which 24 patients were treated with either surgical bypass (n=19), angioplasty (n=1), stent placement (n=2), and medical therapy (n=2). The patients presented with distal emboli (n=3), ischemic ulcer (n=2, one with claudication and ulcers), claudication (n=8), rupture (n=1), aneurysm formation (n=1), pain (back, iliac, inguinal, or abdominal, n=6), acute dissection (n=1), and asymptomatic (n=2). The majority of the patients have been treated with angioplasty. The use of stent placement has been reserved for patients who have failed angioplasty or with refractory stenosis or flow limiting dissection (2, 17).

In the present study, 14 patients with FMD involving the iliac arteries were identified. Some of the risk factors for atherosclerotic vascular disease including diabetes and heart disease were not very common in this series. Hypertension, however, was seen in almost 80% of patients due to concomitant involvement of the renal arteries with FMD in the majority of the patients (86%).

Iliac FMD is usually an asymptomatic disorder (15). Of the 14 patients in this series, only six were symptomatic and all presented with either unilateral or bilateral lower extremity claudication that was treated with angioplasty with or without stent placement. None of the patients presented with critical limb ischemia, peripheral embolism or dissection. Clinical success defined as relief from presenting symptoms at 12 months after endovascular intervention was achieved in all but one patient. One patient did not experience any relief from bilateral lower extremity pain despite technically successful endovascular intervention and normalization of post intervention ABIs. The etiology of this patient's lower extremity pain was later determined to be neurogenic after evaluation by multiple subspecialty services.

This study has some limitations. It is small and is retrospective and does not evaluate the true incidence of this disease. Furthermore, we do not have histology to confirm iliac FMD in our subjects.

In conclusion, FMD of the iliac arteries is relatively rare and its true incidence is unknown. As noted in our series, most but not all patients with iliac FMD have coexisting renal FMD. Although usually asymptomatic, iliac artery FMD can cause limb symptoms by luminal narrowing, microthrombi with distal embolization and EIA thrombosis due to dissection(15). Clinical presentation may mimic atherosclerosis and treatment should be tailored to presenting clinical symptoms. Angioplasty is the preferred treatment of choice and carries a high technical success rate. Endovascular stenting should be considered when angioplasty alone does not improve the hemodynamic characteristics.

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Fig. 1.
Angiographic image of a study subject showing FMD of bilateral external iliac arteries

Table 1

Demographics and clinical characteristics of the patients with iliac artery FMD

	N or mean for continuous variables	Percentage
Demographics		
Number of Patients	14	100%
Age at diagnosis (years)	56 ± 12	
Race (Caucasian)	14	100%
Gender (Women)	12	86%
Cardiovascular risk factors		
Hypertension	11	79%
Diabetes	0	0%
Smoking history	5	36%
Hyperlipidemia	9	64%
Coronary artery disease	3	21%
Chronic Renal Insufficiency	0	0%
Obesity	5	36%
Cerebrovascular Disease	3	21%
Presenting Symptoms		
Claudication/Lower extremity pain	6	43%
Asymptomatic/Incidental	8	57%
Dissection	0	0%
Critical Limb Ischemia	0	0%
FMD of other vascular beds on imaging		
Carotid	5	36%
Coronary	1	7%
Renal	12	86%
Mesenteric	0	0 %
Other (Peripheral)	2	14%

Table 2

Details of the interventions in patients with iliac artery FMD

Procedural Details and Results	N (%)
# of Procedures	10
Access	
RCFA	6 (60%)
LCFA	3 (30%)
Angioplasty alone	8 (80%)
Bilateral EIA	5 (50%)
Right EIA	1 (10%)
Left EIA	2 (20%)
Stent	2 (20%)
Right EIA	1 (10%)
Left EIA	1 (10%)
Technical Success	10 (100%)
Complications	0 (0%)
Clinical Success	7 (70 %)
Symptom Resolution	8 (80%)
Symptom Free Survival (months)	0–153 (Mean = 56.3)

RCFA= Right common femoral artery

LCFA= Left common femoral artery

EIA= Right external iliac artery

Table 3

Details of previous reports on patients with FMD involving iliac arteries

Study	Year	Age, Gender	Presenting Symptoms	Treatment	Artery Involvement
Burri et al	1983	45, M	Inguinal pain	iliofemoral bypass	Left external iliac
Parnell et al	1988			Percutaneous transluminal angioplasty	External iliac
Sauer et al	1990	67, M	Left distal extremity microemboli	Left external iliac resection and Dacron graft	Left external iliac
	1990	55, M	Distal extremity microemboli	Resection with end to end anastomosis	Bilateral external iliac
	1990	71, M	Left distal extremity microemboli	Left external iliac resection and Dacron graft	Left external iliac
	1990	56, F	Acute dissection Presenting with right thigh and leg pain	Conservative with aspirin and exercise	Right external iliac
	1990	62, F	Left thigh and calf claudication	Open surgery with dilatation	Bilateral external iliac
	1990	45, W	Bilateral claudication	Open surgery with dilatation	Bilateral external iliac
	1990	61, W	Left calf claudication and left malleolar ulcer	Aortobifemoral bypass	Bilateral external iliac
	1990	35, W	Bilateral claudication	Open surgery with dilatation	Bilateral external iliac
Patel et al	1990	39, M	Left inguinal pain	Bypass	Left external iliac
Thevenet et al	1992	45, F	Lower limb acute ischemia	Aortobifemoral bypass	Right external iliac
		51, M	Intermittent claudication	Bilateral iliofemoral bypass	Bilateral external iliac
		47, F	Abdominal pain	Bypass	Right external iliac
		29, F	Iliac fossa pain	Interposition with vein	Left external iliac
		53, M	Intermittent claudication	Iliofemoral bypass	Right external iliac
Luck et al	2002	45, M	Intermittent Claudication	Thromboendarterectomy	Right external iliac
Honjo et al	2004	30, M	Shock due to rupture	Iliofemoral bypass	Left common iliac
Asuta et al	2003	69, F	Right common iliac aneurysm	Surgical resection and reconstruction with a Y graft	Right common iliac and left internal iliac
Chin et al	2009	50, F	Asymptomatic	Conservative therapy	Bilateral common iliac
Akashi et al	2010	49, M	Right leg claudication, right lower quadrant abdominal pain	resection and Dacron graft	Right common and external iliac
Okazaki et al	2011	63, F	Left leg claudication	Surgical resection and reconstruction with a Y graft	Bilateral common iliac
		37, M	Sudden on-set back pain	Resection and interposition of bilateral external iliac with grafts	Right external iliac
Rastogi et al	2012	88, F	Right posterior ankle ulcer	Percutaneous transluminal angioplasty with stenting	Bilateral external iliac
Present Series	2013	14 Cases	Asymptomatic (8), Claudication (6)	Percutaneous transluminal angioplasty with or without stenting	Details in Table 2