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Endovascular Therapy for Fibromuscular Dysplasia of the Bilateral External Iliac Arteries Visualized with Optical Coherence Tomography

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF 1 **Takeshi Niizeki**
CD 1 **Mitsunori Ishino**
CDF 1 **Tatsuro Kitahara**
CD 1 **So Yamauchi**
CDE 1 **Eiichiro Ikeno**
AEF 2 **Isao Kubota**

1 Department of Cardiology, Okitama Public General Hospital, Kawanishi, Yamagata, Japan
2 Department of Cardiology, Pulmonology, and Nephrology, Yamagata University School of Medicine, Yamagata, Japan

Corresponding Author: Takeshi Niizeki, e-mail: takeshi.niizeki@okitama-hp.or.jp
Conflict of interest: None declared

Patient: Female, 60
Final Diagnosis: Fibromuscular dysplasia of the bilateral external iliac arteries
Symptoms: Intermittent claudication of the bilateral legs
Medication: —
Clinical Procedure: Endovascular therapy
Specialty: Cardiology

Objective: Rare disease
Background: Fibromuscular dysplasia (FMD) is a non-inflammatory, non-atherosclerotic, degenerative vascular disease that most frequently affects renal and carotid arteries in women aged 30–50 years, and rarely complicating arteries of the lower limbs.

Case Report: A 60-year-old woman was admitted with intermittent claudication of both legs. We performed pelvic and bilateral lower-extremities angiography, which revealed that the bilateral external iliac arteries (EIAs) had the 'string of beads' appearance with a diagnosis of FMD. Endovascular therapy (EVT) was performed for the bilateral EIAs. Optical coherence tomography (OCT) images showed thickening and thinning of the middle layer, while three-dimensional OCT images showed a 'hausta coli'-like appearance. After successful balloon angioplasty, claudication completely disappeared.

Conclusions: We report a rare case of EVT successfully performed for FMD of the bilateral EIAs. Our findings suggest that OCT may provide unique diagnostic clues in FMD patients.

MeSH Keywords: Endovascular Procedures • Fibromuscular Dysplasia • Iliac Artery • Tomography, Optical Coherence

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Background

Fibromuscular dysplasia (FMD) is a non-inflammatory, non-atherosclerotic, degenerative vascular disease that most frequently affects renal and carotid arteries in women aged 30–50 years, rarely complicating arteries of the lower limbs [1,2]. Medial fibroplasia is the most common histologic type of FMD. This wall weakness may be responsible for the formation of intraluminal webs causing stenosis, dissection, and aneurysmal formation. FMD of lower extremities arteries is an uncommon disease of unknown detailed etiology. The literature on FMD of the iliac artery in the past 30 years is almost all single case reports or small case series [3,4]. Recently, the clinical features, presenting symptoms, and vascular events for FMD were reported in multicenter registry data from the United States [5]. Although little is known about optical coherence tomography (OCT) findings and its importance in FMD patients, we report here our experience of successful endovascular therapy (EVT) and OCT findings for FMD of the bilateral external iliac arteries (EIAs).

Case Report

A 60-year-old female was referred to our hospital complaining of intermittent claudication in the bilateral calf, with a walking distance of about 300 meters. Her only coronary risk factor was dyslipidemia. Cardiac examination revealed a regular rhythm with no murmurs. A routine hemogram and biochemistry data were within normal limits. In addition, eosinophil

Figure 1. Digital subtraction angiography showed that the bilateral external iliac arteries had stenosis with the 'string of beads' appearance consistent with fibromuscular dysplasia.

cell count was not elevated. She had no history of connective tissue disease, which was also excluded by physical and laboratory examinations. Her ankle-brachial index (ABI) was 0.72 on the right leg and 0.77 on the left, and peripheral artery disease was suspected. As shown in Figure 1, digital subtraction angiography of the bilateral lower-extremities showed that the bilateral EIAs had stenosis with the 'string of beads' appearance consistent with multifocal FMD. Savard et al. reported the angiographic classification of FMD; FMD lesions were

Figure 2. Intravascular ultrasound revealed the non-atherosclerotic lesion and the formation of intraluminal webs causing stenosis.

Figure 3. Optical coherence tomography images clearly showed thickening and thinning of the middle layer (A), while three-dimensional Optical coherence tomography images showed a 'haustra coli'-like appearance (B).

classified as multifocal FMD if there were at least 2 stenoses in the same arterial segment, otherwise they were classified as unifocal [6].

Based on these angiograms, we offered her a low-invasive approach using endovascular therapy (EVT) for the bilateral EIAs. We prepared bi-common femoral arteries with the 6Fr system Zemex sheath 25 cm (Zeon Medical Corporation, Tokyo, Japan) inserted from bi-common femoral arteries. An attempt to pass the stenotic site by use of a 0.014-inch chevalier floppy wire (Future Medical Design Corporation, Saitama, Japan) was performed, and the guide wire easily crossed. There were 40-50 mmHg pressure gradients in the bilateral EIAs lesions. Intravascular ultrasound (IVUS) (ViewIT, Terumo Corporation, Tokyo, Japan) revealed the non-atherosclerotic lesion and the formation of intraluminal webs causing stenosis (Figure 2). Furthermore, OCT images (C7 XR OCT Intravascular Imaging System, St. Jude Medical, Minnesota, USA) clearly showed thickening and thinning of the middle layer (Figure 3A), while three-dimensional OCT images showed a 'haustra coli'-like appearance (Figure 3B). Thus, we considered that IVUS and OCT may be used as an adjunctive tool to diagnose and evaluate

FMD. We performed balloon angioplasty using a 6.0×80 mm balloon [Jackal RX 6.0×80 mm (Kaneka Corporation, Osaka, Japan)]. After angioplasty, pressure gradients in the bilateral EIAs lesions were decreased to 10–15 mmHg. Because it has been reported that patients with iliac artery FMD have coexisting FMD of the renal and coronary arteries [3], we performed renal and coronary angiography. However, there were no abnormal findings on the renal and coronary arteries in this case. The sheaths were removed, and the patient was discharged on the following day. The post-operative course was uneventful. The follow-up ABI at 4 weeks was improved (0.92 in the right leg and 0.91 in the left) and clinical success with symptom resolution was achieved. Balloon angioplasty alone had higher restenosis rates than angioplasty with stent. Although conventional balloon angioplasty without stenting appeared successful in this case and other reports about FMD [3–6], close follow-up care is prudent for possible restenosis by interview and ABI measurement at 1, 3, 6, and 12 months after EVT, and every 6 months thereafter. In addition, there were no abnormal findings on the cerebral and carotid arteries with magnetic resonance imaging in this patient.

Discussion

Here we report a rare case of EVT for FMD of the bilateral EIAs successfully performed with support of imaging devices (IVUS and OCT). FMD mostly occurs in the renal artery (70%), followed by the carotid artery (25%) [1,2]. Because stenotic lesions in lower extremities arteries are mostly caused by arteriosclerosis obliterans, FMD of the EIA as in this case is extremely rare. It has been reported that the incidence of iliac artery FMD in patients with renal and/or carotid artery FMD is about 5% [4]. Although the exact prevalence of FMD in the lower extremities arteries is unknown, EIA appears to most commonly involve the lower limbs [3,4]. It is important to elucidate the prevalence, natural history, optimal methods of diagnosis, and treatment of FMD by a multicenter registry in the future.

Although angiography remains the gold standard imaging technique to diagnose and evaluate FMD for lacking specific markers or effective non-invasive tests, it cannot accurately determine the degree of arterial stenosis. The recent introduction of OCT is a novel method for obtaining tomographic images of a human organ based on the coherence of light. The resolution of OCT (to 10–20 μm) is about 10-fold higher than that of IVUS (to 100–150 μm) [7]. Thus, OCT can differentiate the internal and external laminae, and is superior ability of detecting web formation in FMD than IVUS as shown Figures 2 and 3. Catheter-based angiography was useful and excellent for diagnostic confirmation of FMD. However, if the diagnosis, the degree of stenosis, and therapeutic effect remained uncertainly,

OCT findings might adjunctive tools. Thus, we considered that OCT might be a useful and feasible method for evaluating the obstructive webs and may aid in identifying the FMD. Recently, histopathology was rarely available owing to the infrequency of open surgery; therefore, collecting OCT data with angiography from a large number of patients with FMD might help to elucidate the pathophysiology.

Recently, EVT has been widely applied to aorto-iliac artery disease [8] and a few successful EVT cases for FMD of the extremities arteries have been reported [9,10]. The treatment of symptomatic obstructive FMD mainly consists of EVT without stent, because balloon angioplasty has been mostly shown to sufficiently decrease the pressure gradient without vessel injury [1,2,5,6]. However, stent placement should be reserved for failed balloon angioplasty cases with refractory stenosis or flow-limiting dissection [3,5,6]. In conclusion, FMD should be considered as a potential cause of intermittent claudication in middle-aged women. We consider that balloon angioplasty for FMD of the iliac artery supported by OCT device is a suitable treatment and has a high technical success rate.

Conclusions

We reported a rare case that EVT was successfully performed for FMD of the bilateral EIAs. Our findings suggest that OCT may provide unique diagnostic clues in FMD patients.

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