Case Report

Abdominal Aortic Aneurysm Caused by Aortic Fibromuscular Dysplasia: A Case Report

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Fibromuscular dysplasia (FMD) mainly develops in medium-sized arteries, including renal, extracranial, and extremity arteries, but it rarely causes abdominal aortic aneurysm (AAA). A 69-year-old woman with AAA diagnosed on ultrasonography by a home doctor visited our hospital. Contrast-enhanced computed tomography revealed a saccular aneurysm of terminal abdominal aorta. We performed abdominal aortic replacement and resected the section with aneurysm. Pathological examination of the wall tissue of the resected aneurysm revealed findings that are consistent with FMD. We report this case of AAA caused by aortic FMD because of its rarity.

Keywords: fibromuscular dysplasia, abdominal aortic aneurysm, aortic fibromuscular dysplasia

Introduction

Fibromuscular dysplasia (FMD) is an idiopathic, segmental, nonatherosclerotic, noninflammatory vascular disease that mainly develops in medium-sized arteries and is known to cause renovascular hypertension1) and may lead to stenosis, occlusion, dissection, and/or aneurysm. The clinical presentation of FMD varies with vascular distribution, e.g., the involvement of renal arteries, carotid arteries, and extremity arteries can present as renovascular hypertension, stroke, and limb ischemia, respectively. Other cases of FMD may be asymptomatic and incidentally discovered by imaging.2) Most previous reports on FMD focused on the development of the condition in renal arteries, extracranial carotid arteries, vertebral arteries, or lower extremity arteries.1–3) Conversely, FMD in the abdominal aorta is very rare. Some cases of aortic FMD exist in the literature; however, most of these cases are related to coarctation or dissection, and cases of aortic aneurysm are extremely rare.3,4) We report a case of abdominal aortic aneurysm (AAA) caused by aortic FMD.

Case Report

A 69-year-old woman was diagnosed with AAA by a home doctor; ultrasonography revealed that the AAA was 30 mm in diameter. Three years later, she was referred to our hospital because the AAA increased to 36 mm in diameter. She had hypertension and dyslipidemia. She had no history of smoking or open abdominal surgery. There was no family history of FMD, aortic disease, connective tissue disease, or sudden death.

On physical examination, she was 152-cm tall and weighed 57 kg, without any features of Marfan, Ehlers–Danlos, or Loeys–Dietz syndrome. Her blood pressure was 146/84 mmHg on both arms. Her ankle–brachial index was 1.05 on the right lower limb and 1.01 on the left lower limb. Blood examination was almost normal. Enhanced computed tomography (CT) revealed a saccular aneurysm of terminal abdominal aorta and narrow infrarenal abdominal aorta (10 mm), as well as bilateral common iliac arteries (6 mm) (Fig. 1). The thoracic aorta, suprarenal abdominal aorta, celiac artery, superior mesenteric artery, and bilateral renal arteries appeared normal. Coronary artery stenosis or aneurysm were absent on coronary angiography.

We planned a surgery to address the saccular aneurysm. Due to the narrow diameters of the abdominal aorta and common iliac arteries, even the minimum sized stent-graft available in Japan was more than twice the diameters of her aorta and common iliac artery. Hence, we considered that it is difficult to treat her with endovascular aortic repair (EVAR). Since her aortic aneurysm had a specific shape and she was younger than most patients with AAA,
we decided to perform an open surgery. During the operation, the aneurysm was located at the terminal abdominal aorta and there was no periarterial inflammation, atherosclerosis, or stenosis (Fig. 2A). We performed abdominal aortic replacement with the standard surgical procedure using a 12 mm × 6 mm woven graft (HEMASHIELD PLATINUM, Maquet, Getinge Group Japan K.K., Tokyo, Japan) and resected a section of the aneurysmal wall tissue for pathological examination (Fig. 2B). The postoperative course was good and repeated enhanced CT revealed no abnormalities in the reconstructed section and other arteries. She was discharged from our hospital on postoperative day 13. At a 1-year follow up, she was doing well without recurrence of aneurysm or any symptoms.

Pathologic examination of the aneurysmal wall revealed fragmented elastic fibers in the inner media (Fig. 3), which were almost replaced by collagen fibers with coexisting hyperplastic smooth muscle cell bundle in the outer media. There were no findings of atherosclerosis, denaturation, inflammation, or infection. Furthermore, the aortic wall
had thinned out. Overall, the histologic findings were similar to those of FMD. In addition, because of the absence of other etiologic presentations that predispose the patient to aortic aneurysm development, we diagnosed aortic aneurysm caused by aortic FMD.

**Discussion**

FMD is a rare disorder of medium-sized muscular arteries, including the renal and carotid arteries. There have been few previous reports on aortic FMD. In the report by Olin et al., 447 patients were diagnosed with FMD based on imaging and clinical features while aortic aneurysm was noted in 15 patients (3.4%) with FMD in various arteries, but there were no cases of aortic FMD. Almost all previously reported cases of aortic FMD were diagnosed on the basis of histopathological examination. Tasaki et al. reported that there were 33 cases of aortic FMD in English literature, 17 cases of which were in patients older than 18 years. Aortic FMD cases in patients younger than 18 years were complications of congenital aortic coarctation. In the 17 patients with aortic FMD, coarctation occurred in 12 cases (71%), dissection in 4 cases (23%), and aneurysm (excluding postcoarctation dilatation or dissection) in 1 case. Only 18 cases of aortic FMD, including our case, have been reported and only 2 of these cases were AAAs. Of these 18 cases, 15 (including our case) were diagnosed pathologically.

Characteristic histopathological findings of FMD are loss of elastic fibers, collagen deposition, and proliferation of smooth muscle cells and/or myofibroblasts leading to architectural disorganization in the arterial wall. In 1971, Harrison and McCormack histologically classified FMD under renal arterial disease in renovascular hypertension and this classification was later adopted for the other arteries. Based on the arterial wall layer (intima, media, and adventitia), in which the lesion predominates, FMD was classified into 3 categories. Medial FMD, which is the most common histologic variant, is divided into subgroups of medial fibroplasia, perimedial fibroplasia, and medial hyperplasia. However, because this classification was developed for muscular arteries, it may not be appropriate to use for an elastic artery such as the aorta. The pathological diagnostic criteria of aortic FMD have not been established because of the small number of cases. In previous cases, they were diagnosed with aortic FMD because they had typical histological findings similar to FMD and did not present findings of other aortic diseases such as atherosclerosis, denaturation, inflammation, or infection. In our case, it was not difficult to distinguish between atherosclerosis, denaturation, inflammation, or infection. Initially, we thought that the pathological findings in this case were similar to segmental arterial mediolysis (SAM); however, we finally diagnosed our case with FMD rather than SAM because the smooth muscle cell vacuolization and intact tunica media in the form of islands were absent as the typical findings of the pathological diagnosis.

To date, the clinical diagnostic criteria for aortic FMD awaits establishment. The treatment of an aneurysm with a stent-graft does not reveal whether an aneurysm was caused by aortic FMD or not. We consider that AAA caused by aortic FMD should be suspected without pathological examination before surgery. However, the typical image findings of AAA caused by aortic FMD remain unclear because of their rarity. Our case suggested the narrow infrarenal aorta and the saccular aneurysm protruding anteriorly. In addition, a prior report presenting AAA caused by aortic FMD suggested the saccular aneurysm protruding anteriorly. The infrarenal aorta in the case was narrow and tortuous and appeared like a “string of beads” appearance, which is typical of medial fibroplasia. Although our case did not present the “string of beads” appearance, the narrow renal arteriolar aorta and the saccular aneurysm protruding anteriorly are common in 2 cases. Reportedly, medial fibroplasia causes arterial stenosis and poststenotic dilation, displaying the “string of beads” appearance. In the 2 cases of AAA caused by FMD, these aneurysms might have been caused by poststenotic dilation.

The treatment for FMD differs with regard to the presence of symptoms, location, and type of lesion (i.e., stenosis, dissection, or aneurysm). For symptomatic FMD cases, including renal artery FMD with renovascular hypertension, endovascular revascularization or surgical treatment have been performed. Most treatment decisions in patients with FMD were based on data derived from case reports or small retrospective case series and there is no consensus on the therapeutic indications for asymptomatic FMD, including aneurysms. The treatment for aneurysm is mainly aimed at preventing rupture. Recently, EVAR for the treatment of AAA has increased, but the efficacy of stent-graft therapy for aortic FMD is uncertain because of the absence of previous reports and consensus. In our case, we considered that surgery was better than endovascular treatment because of the irregularly shaped saccular aneurysms and narrow infrarenal aorta. The efficacy and safety of endovascular treatment for an atypical aneurysm (such as FMD, infected aneurysm, or connective tissue disease) have not been elucidated. Some studies have reported the treatment of FMD and renal artery aneurysm with a stent graft. While this procedure is useful and minimally invasive, its long-term outcomes remain unknown. In addition, if the endovascular treatment for FMD increases, the opportunity for the pathological analysis of FMD could decrease. In our opinion,
surgical treatment of AAA caused by FMD is better than the endovascular treatment until sufficient clinical and pathological findings are obtained.

Conclusion

We reported a very rare case of AAA caused by aortic FMD. To establish pathological/clinical diagnostic criteria and treatment strategy (including endovascular treatment) for aortic FMD, further accumulation of pathologic data and clinical experience is necessary. Considering the rarity of this disease, the number of cases in a single center is not adequate for establishing diagnostic and treatment criteria. More reports from multiple institutions are necessary.

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Disclosure Statement

All authors have no conflict of interest to disclose.

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Manuscript writing: RK
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