

[ CASE REPORT ]

## Sequential Symptomatic Arterial Dissections in Multiple Vascular Beds in a Patient with Fibromuscular Dysplasia

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### Abstract:

A 60-year-old man was admitted to our hospital because of abdominal pain and disturbed consciousness. Head magnetic resonance imaging showed right vertebral artery dissection and abdominal enhanced computed tomography showed dissection of the superior mesenteric artery. The patient was diagnosed as having fibromuscular dysplasia (FMD) based on conventional angiography. Although multiple vascular bed involvement is observed in approximately 40% of FMD patients, reports of sequential symptomatic dissections in various vascular beds are rare. Patients with FMD and dissection require close observation, and hemodynamic stabilization may prevent not only the further development of dissection, but also subsequent dissection of other arteries.

**Key words:** fibromuscular dysplasia, ischemic stroke, superior mesenteric artery, dissection, multiple vascular beds

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### Introduction

Fibromuscular dysplasia (FMD) is a non-atherosclerotic, non-inflammatory vascular disease and multiple vascular bed involvement has been observed in around 40% of FMD patients. However, reports of simultaneous symptomatic dissection in various vascular beds are scarce. Herein, a FMD patient presenting simultaneous arterial dissection in multiple vascular beds, is reported.

### Case Report

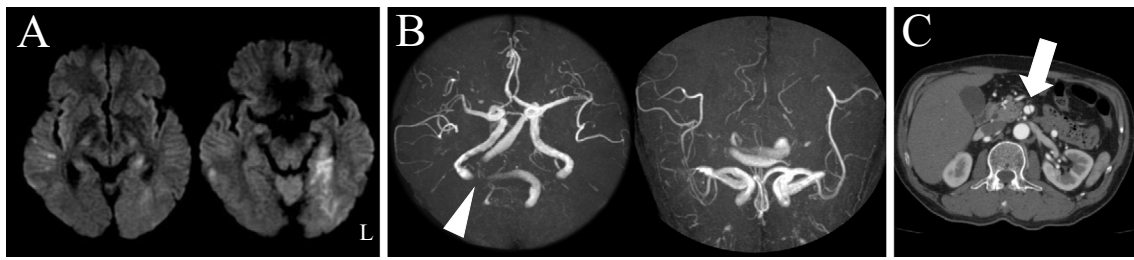
A 60-year-old right-handed man was transferred to our hospital because of abdominal pain and disturbed consciousness. He initially complained of abdominal pain, and then his consciousness became disturbed approximately 2 hours later. He had suffered an ischemic stroke 12 years previously, and an intracranial aneurysm of the anterior cerebral artery was found at that time. Although he was taking a single antihypertensive medication due to the presence of the

intracranial aneurysm, he had never diagnosed with hypertension. He had no family history of connective tissue diseases such as Ehlers-Danlos syndrome or Marfan syndrome. On arrival, his blood pressure (BP) was 166/110 mmHg, and his heart rate was 76 beats/min with a regular rhythm. A general physical examination showed epigastric tenderness without signs of peritoneal irritation. A *Treponema pallidum* hemagglutination (TPHA) test was negative for syphilis. A neurological examination revealed consciousness disturbance (Glasgow Coma Scale: E3, V3, M5). Magnetic resonance imaging (MRI) on admission showed hyperintense lesions in the bilateral posterior cerebral artery (PCA) territories on diffusion-weighted imaging (Fig. 1A), and the signal intensities of the right vertebral artery (VA), lower basilar artery (BA), and proximal bilateral PCAs were decreased on MR angiography (MRA, Fig. 1B, arrowhead). Enhanced computed tomography (CT) showed dissection of the superior mesenteric artery (SMA, Fig. 1C, arrow). Beading was not observed in the SMA, and with the exception of the cervical arteries and the SMA, no arteries showed abnormalities on enhanced CT.

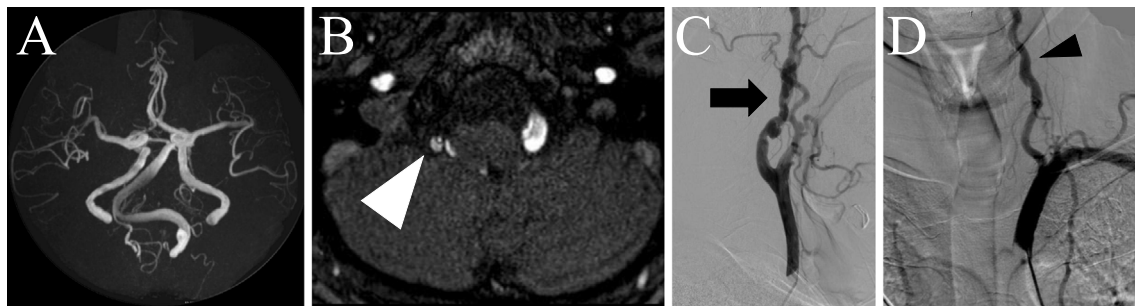
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**Figure 1.** Magnetic resonance imaging and enhanced computed tomography on admission. Diffusion-weighted imaging shows hyperintense lesions in the bilateral posterior cerebral artery territories (A), and magnetic resonance angiography shows the diminished signal intensity of the right vertebral artery, lower basilar artery (arrowhead), and proximal bilateral posterior cerebral arteries (B). Enhanced computed tomography shows dissection of the superior mesenteric artery (C, arrow).



**Figure 2.** Follow-up magnetic resonance imaging and conventional angiography. Magnetic resonance angiography (MRA) on day 2 shows re-visualization of the right vertebral artery (VA) and basilar artery (A). The MRA source image on day 6 shows a double lumen and intramural hematoma in the right VA (B, white arrowhead). Conventional angiography shows alternating stenosis and dilatation in the middle cervical portion of the internal carotid artery (C, black arrow) and tortuous change in the V2 portion of the left VA (D, black arrowhead), with no atherosclerotic lesions at the origin of each artery.

The disturbed consciousness improved after fluid replacement therapy, and the abdominal pain gradually subsided without symptoms of intestinal ischemia. Follow-up MRI on day 2 showed the re-visualization of the right VA and BA (Fig. 2A). MRI on day 6 demonstrated a double lumen and intramural hematoma in the right VA (Fig. 2B, arrowhead). Conventional angiography showed alternating stenosis and dilatation in the middle cervical portions of the bilateral internal carotid arteries (ICAs) and tortuous change in the V2 portion of the left VA, with no atherosclerotic lesions at the origin of each artery (Fig. 2C and D). The patient was diagnosed as having right VA and SMA dissection due to FMD, based on the extracranial ICA “string-of-beads” sign without atherosclerotic change, spontaneous SMA dissection, and intracranial aneurysm (1), as well as the negative TPHA test result and the absence of a family history of connective tissue disease. The patient was left with only a slight reading disturbance as a neurological sequela. At the time of writing, the patient has been receiving conservative treatment for 4 months; there has been no recurrence of stroke or any abdominal symptoms during the follow-up period.

## Discussion

In the present patient with FMD, it is possible that the first arterial dissection occurred in the SMA and that this was followed by a second dissection in the right VA. The sudden-onset epigastric pain, enhanced CT findings, and serial MRA changes suggested acute SMA and right VA dissections. FMD is a non-atherosclerotic, non-inflammatory vascular disease (2), and although multiple vascular bed involvement is observed in approximately 40% of FMD patients (3), reports of sequential symptomatic dissections in different vascular beds are rare (4).

Although the exact timing of the onset of each dissection (SMA and VA) was unclear, the SMA dissection was assumed to have preceded the VA dissection in the present case because epigastric pain preceded the disturbance of consciousness. The precise mechanisms of the sequential dissections in the present case are unknown; however, changes in hemodynamics induced by the SMA dissection itself or epigastric pain may have played an important role: high BP promoted by dissection or pain could result in dissection of the right VA. FMD patients presenting with arte-

rial dissection may be prone to tears of other arteries, even in other vascular beds. Patients with FMD and dissection require close observation, and hemodynamic stabilization may prevent not only further dissection, but also the subsequent dissection of other arteries.

**The authors state that they have no Conflict of Interest (COI).**

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