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Fibromuscular Dysplasia Presented With Spontaneous Dissection of the Left Main Artery

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A 31-year-old woman with a history of trisomy 10, subarachnoid hemorrhage, and previous myocardial infarction presented with asystolic cardiac arrest preceded by chest pain and shortness of breath. After return of spontaneous circulation, electrocardiogram showed ST-segment elevation in the anteroseptal leads. Cardiac biomarkers were elevated. Coronary angiogram showed spontaneous dissection of the left main artery extending into proximal left anterior descending and left circumflex arteries. The mid segment of the left anterior descending artery had an aneurysm, and the obtuse marginal artery showed smooth narrowing of the distal segment (**Figure 1**, Online Video 1). The right coronary artery was normal. An intra-aortic balloon pump was placed for hemodynamic support, but the patient died during the cardiac catheterization after multiple episodes of asystolic cardiac arrest. Coronary fibromuscular dysplasia (FMD) was suspected and confirmed by the finding of beading of the cavernous internal carotid arteries on a computed tomography angiogram of the head obtained 8 years before (**Figure 2**).

An FMD is an arteriopathy that predominantly affects middle-aged women (1). The etiology of FMD is still unknown, although genetic predisposition is likely. The renal and cervicocranial arteries are most commonly involved, while the coronary arteries are rarely affected in FMD (2). The classic angiographic "sting of beads" that may be observed in the renal artery rarely occurs in coronary arteries. Spontaneous coronary arteries dissection (SCAD) is the most common manifestation of coronary FMD; FMD presents in most patients with SCAD (1). Other angiographic findings include aneurysmal dilation, smooth narrowing, or distal tapering typically with intramural hematoma, spasm, and tortuosity (2). SCAD may be initiated by an intimal tear that leads to propagating medial dissection, or by a dissecting medial hematoma, initiated by rupture of vaso vasorum (1). In patients in hemodynamically stable condition with SCAD, conservative therapy, with aspirin and beta blockers, is preferred. Patients with symptoms of ongoing ischemia or hemodynamic compromise should be considered for revascularization, which can be technically challenging and associated with higher complications (3).

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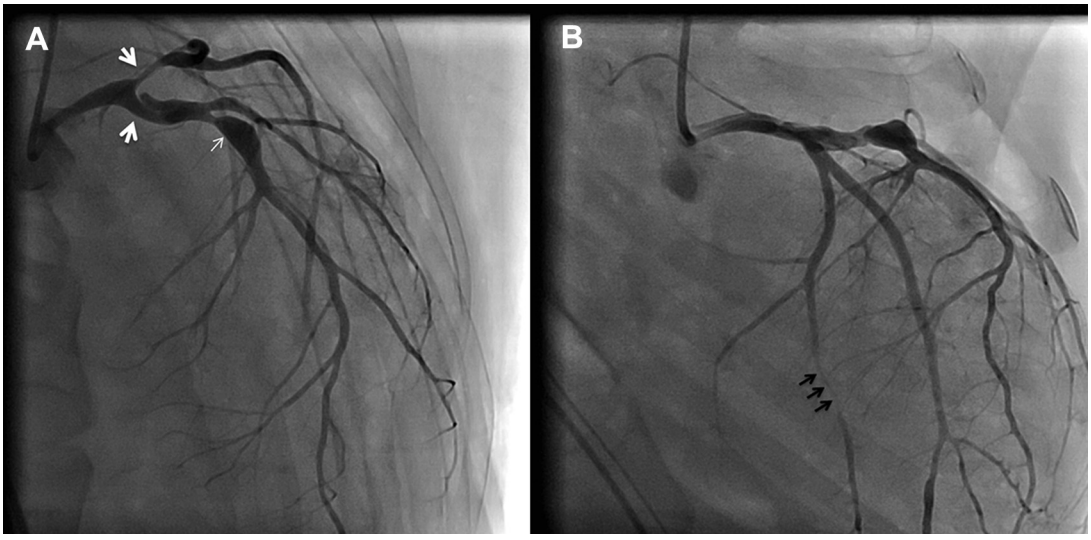
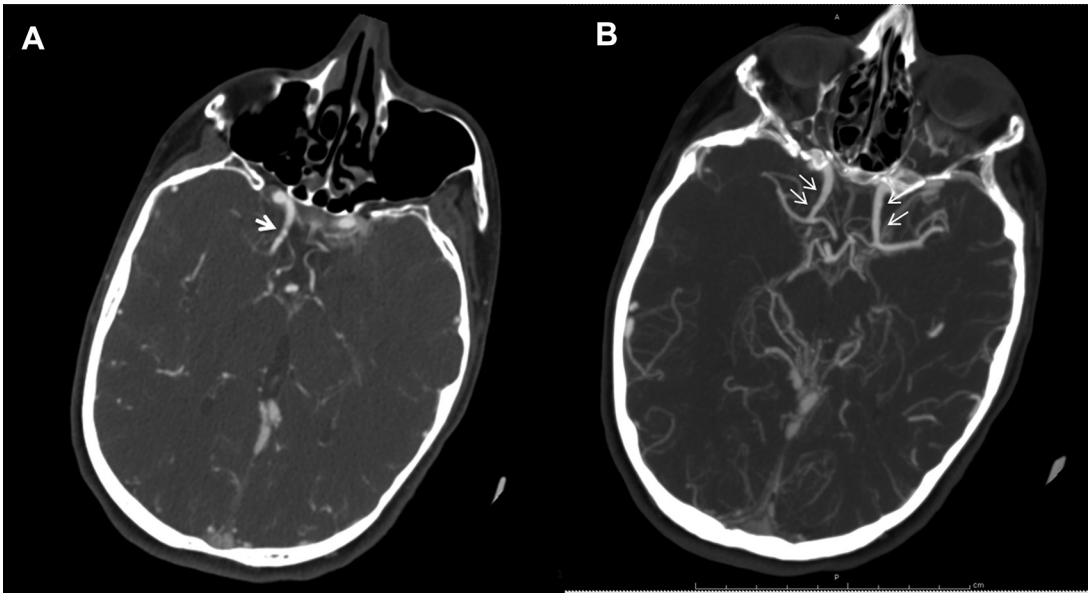
FIGURE 1 Left Coronary Angiography



Left coronary angiography showing **(A)** dissection of the left main artery (Online Video 1) extending into proximal left anterior descending and left circumflex arteries **(white thick arrows)**. The mid segment of the left anterior descending artery had an aneurysmal dilatation **(white thin arrow)**, **(B)** The obtuse marginal artery showed smooth narrowing of the distal segment **(black arrows)**.

FIGURE 2 Computed Tomography Angiogram of the Head



Computed tomography angiogram of the head showing **(A)** mild beading of the right cavernous internal carotid artery **(arrow)**. **(B)** Computed tomography angiogram of the head, reconstruction image, showing mild beading of both right and left cavernous internal carotid arteries **(arrows)**.

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KEY WORDS coronary artery disease, chromosome 10, fibromuscular dysplasia, spontaneous coronary artery dissection, trisomy 10

APPENDIX For a supplemental video and its legend, please see the online version of this article.