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Six-Vessel Spontaneous Coronary Artery Dissection and Vertebral Artery Dissection in Fibromuscular Dysplasia

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Abstract Fibromuscular dysplasia is a non-inflammatory and non-atherosclerotic arteriopathy commonly affecting medium-size renal, carotid, and vertebral arteries. Involvement of the coronary artery is very rare but may cause life-threatening spontaneous dissection. Because it is rare, this disease may go unrecognized. A unique angiographic finding of non-coronary arteries can make an early diagnosis, and prompt treatment is paramount. In this paper, we report on a multiple vessel coronary artery dissection in a female patient who presented with acute stroke symptoms secondary to spontaneous dissection of the left vertebral artery. To date, this is the first case report of the spontaneous dissection of six branches of coronary arteries and the vertebral artery due to fibromuscular dysplasia.

Keywords: *spontaneous coronary artery dissection, vertebral artery dissection, fibromuscular dysplasia, coronary angiogram*

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1. Introduction

Fibromuscular dysplasia (FMD) is a non-inflammatory, non-atherosclerotic, medium-size arteriopathy, which commonly affects the renal arteries (79.7%), extra-cranial carotid arteries (74.3%), and vertebral arteries (36.6%) [1]. It has been reported in every arterial territory, including the coronary arteries [1]. FMD predominantly occurs in female patients, with a mean age of 52 years (standard deviation of 13.4 years) at diagnosis [1]. The true prevalence of FMD is unknown, but it is thought to be a very rare clinical entity [1] that is not commonly recognized. In the largest U.S. FMD registry of 447 patients, presenting symptoms that led to the diagnosis of FMD were hypertension (63.8%) and headache (52.4%). Myocardial infarction was only reported in 1.3% [1]. Here, we discuss a case of six-vessel spontaneous coronary artery dissection (SCAD) and left vertebral artery dissection in FMD.

2. Case Presentation

A 34-year-old non-pregnant female presented with right-sided weakness and left facial numbness. An MRI of the head showed an acute ischemic stroke of the posterior aspect of the left medulla, and computed tomography angiography of the head and neck revealed left vertebral artery dissection. The patient was given a diagnosis of acute ischemic stroke from spontaneous dissection of the left vertebral artery (Figure 3). A few hours following a heparin infusion, the patient developed a sudden onset of chest pain and shortness of breath. Her blood pressure was 82/55 mmHg. On physical examination, her lungs revealed crackles in the bases bilaterally. Her heart exam was essentially normal. The chest X-ray showed pulmonary edema. An electrocardiogram showed ST-segment elevation in leads I and aVL with reciprocal ST depression in leads II, III, aVF, and V3-5. The patient's troponin peaked at 89.59 ng/mL (normal: 0-0.01 ng/mL). Autoimmune serology tests were negative. A urine drug screen was negative for cocaine. An echocardiogram revealed severe global hypokinesis and distal anterior wall akinesis of the left ventricle with an ejection fraction of 20-25%.

The patient underwent an emergent coronary angiogram, and it showed a 70% eccentric smooth narrowing of the left main coronary artery (LM) (Figure 1A), a 90% smooth long narrowing and stenosis of the mid-left anterior descending artery (LAD) (Figure 1A-B), and a 90% smooth narrowing, stenosis, and intimal flap of the first diagonal artery (1st D) (Figure 1C). There was an intimal flap of the left circumflex artery (LCx) and a 100% occlusion of the proximal segment of the first obtuse marginal artery (1stOM) (Figure 1B). There was an 80% stenosis of the mid-right coronary artery (RCA), with a mid-long smooth narrowing lesion (Figure 1D). A femoral angiogram was performed before placement of a closure device. It showed a beaded appearance of the right external iliac artery, suggestive of FMD (Figure 2). Because of hemodynamic instability and dissection of the left main

artery and the other five coronary vessels, an emergency coronary artery bypass graft (CABG) was performed. After the CABG, the patient's clinical condition improved. Renal artery duplex showed 0-59% bilateral stenosis.

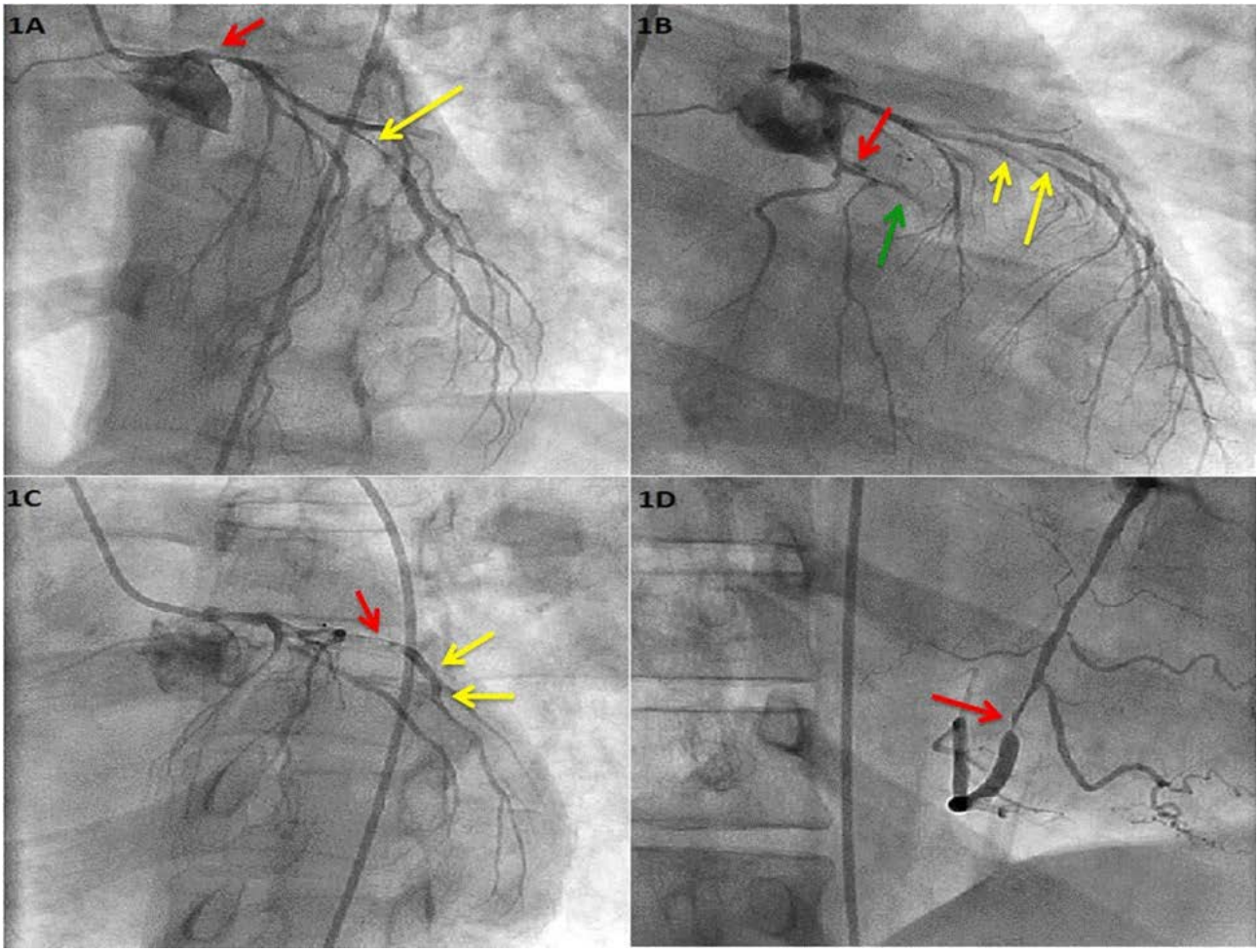

Figure 1. Coronary angiogram: **(A)** Narrowing of the left main coronary artery (red arrow), and long narrowing stenosis of mid left anterior descending artery (yellow arrow). (**B)** Dissection (intimal flap) of left circumflex artery (red arrow), 100% occlusion of proximal segment of the first obtuse marginal artery (green arrow), and long narrowing stenosis of mid left anterior descending artery (yellow arrows). (**C)** Narrowing stenosis (red arrow) and dissection (intimal flap) of the first diagonal artery (yellow arrows). (**D)** Stenosis of mid right coronary artery with mid long smooth narrowing lesion (red arrow)

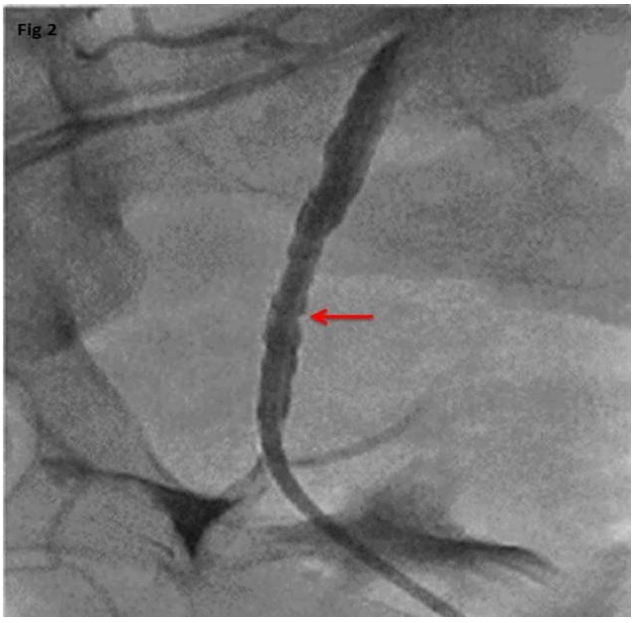

Figure 2. Femoral angiogram: Beaded appearance of the right external iliac artery (red arrow), suggestive of FMD

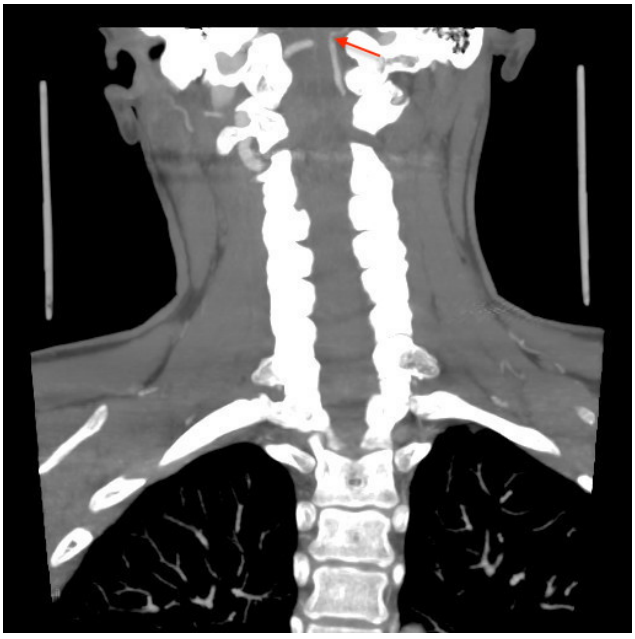

Figure 3. CTA Neck: Coronal view showing irregularity with narrowing of left vertebral artery suggestive of dissection (red arrow)

3. Discussion

FMD is commonly diagnosed by angiographic imaging [1]. A string of beads appearance is associated with medial fibroplasia. This is the most common type of FMD, affecting the renal, carotid, and vertebral arteries [1]. Though a classic beaded appearance supports the diagnosis of FMD, it is not found in coronary vessels [2]. For this reason, the diagnosis of FMD in spontaneous coronary artery dissection (SCAD) depends on the angiographic findings from non-coronary arteries.

SCAD most commonly occurs in the peripartum period. Other risk factors include the diagnosis of Ehler-Danlos or Marfan syndrome, a history of oral contraceptive pills, and FMD [3]. In a retrospective study of 87 patients with angiographic-confirmed SCAD, ten patients (11%) were found to have FMD in their non-coronary arteries. Eight of those patients had an angiographically irregular beaded appearance of FMD in the external iliac artery based upon a femoral angiogram before closure device placement [3]. SCAD is a rare condition found in only 0.7% of FMD patients who experienced coronary artery dissection [1]. To date, there have been only seven cases of triple-vessel coronary artery dissection reported due to FMD [4-10]. This paper presents the first case of six-vessel/branch SCAD with concomitant vertebral artery dissection as a rare, fatal, cardiovascular complication of FMD.

In acute SCAD, optimal management remains undetermined, given the lack of comparative studies. In one retrospective study of 189 patients with SCAD, the patients treated with initial revascularization often presented with ST elevation myocardial infarction, coronary artery occlusion, and higher mean lesion stenosis in the larger coronary arteries based upon coronary angiograms. Percutaneous coronary intervention (PCI) was associated with high rates of emergency CABG due to PCI failure. Patients treated with CABG as the initial therapy had excellent in-hospital outcomes. The conservative group had good in-hospital outcomes; however, 10% developed SCAD progression. There were no differences in rates of myocardial infarction, revascularization, and recurrent SCAD among the revascularization or conservative group at five years [11]. Initial revascularization may be an appropriate strategy for clinically unstable patients or those with low coronary blood flow on angiogram [11]. As illustrated in this case, the patient underwent CABG because of the severity of symptoms and multiple vessel involvement. In patients with multi-vessel spontaneous dissection of the coronary and non-coronary arteries, fibromuscular dysplasia should be included in the differential diagnosis. The classic beaded appearance of the external iliac artery on femoral angiogram adds supportive evidence for this diagnosis.

Conflict of Interest

The authors have no conflict of interest.

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