Kosin Medical Journal 2017;32:240-243.
https://doi.org/10.7180/kmj.2017.32.2.240

KMJ
Case Report

Spontaneous Coronary Artery Dissection in a female patient with fragile X syndrome

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We report a case of Spontaneous coronary artery dissection associated with fragile X syndrome. The relationship between fragile X syndrome and Spontaneous coronary artery dissection is unclear. However, More research will need about the causes and treatment of Spontaneous coronary artery dissection.

Key Words: Acute coronary syndrome, Fragile X syndrome, Spontaneous coronary artery dissection

Spontaneous coronary artery dissection (SCAD) is a rare but significant cause of acute coronary syndrome and sudden cardiac death. The first angiographic report of SCAD was done by Ciraulo in 1978.[1] SCAD may present with a variety of symptoms, such as acute myocardial infarction, ventricular arrhythmiaa, and sudden cardiac death, or may also be asymptomatic.[2]

SCAD is most often diagnosed using coronary angiography. The pathogenesis of SCAD is unclear, but female gender, the peripartum period, and atherosclerosis are known to increase the risk of developing the condition.[3,4] Genetic disease which involve connective tissue such as Marfan syndrome, Ehlers-Danlos syndrome and autoimmune disease such as systemic lupus erythematosus also known to be increased risk of SCAD.[5] However, the relevance of the fragile X syndrome (FXS) has not yet been reported. Here, we report the case of a woman with SCAD and FXS.

CASE

A 45 year old woman visited the emergency room (ER) due to sudden anterior retrosternal chest pain. She did not have any coronary risk factors such as smoking, diabetes, hypertension and dyslipidemia at all. We were informed that she was an FXS carrier by another hospital. Electrocardiography which was taken in ER, revealed marked ST segment elevation in the V2, V3, and V4 leads. Transthoracic echocardiography showed hypokinesia of the anterior segments. Cardiac enzymes levels were within normal ranges at the time of admission.

Coronary angiography revealed a linear coronary

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Received: Oct. 12, 2015
Revised: Oct. 13, 2015
Accepted: Oct. 13, 2015

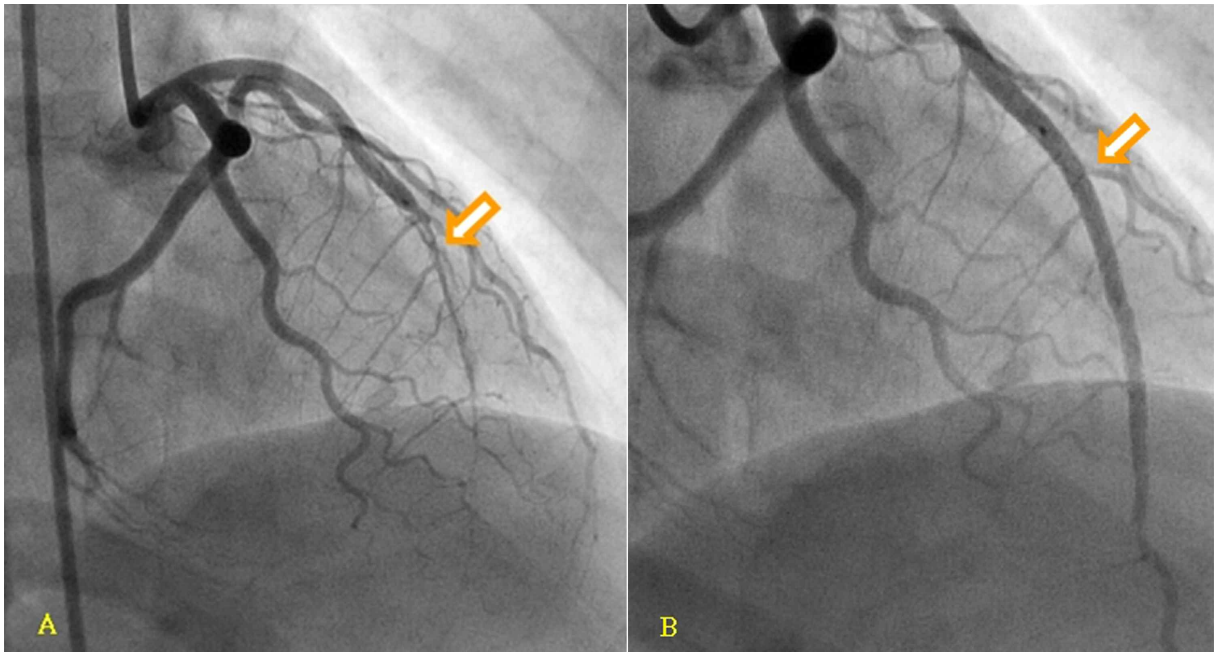

Fig. 1A : Dissection of the left anterior descending artery (arrow), revealed by coronary angiography
1B : Resolution of the intimal dissection after stent insertion (arrow)

artery dissection from the middle-to-distal left anterior descending artery (LAD)(Fig. 1A). Entry site of dissection was not seen but echo-lucent area under intima was seen from the mid left anterior descending aretery (LAD) which was transformed to hematoma distally in intravascular ultrasonography (IVUS)(Fig. 2A). Percutaneous coronary intervention was performed because the continuous chest pain of the patient and persistent ST segment elevation on monitoring ECG. A 2.75 × 26 ㎜ drug-eluting stent (Orsiro 2.75 × 26, upto 8 atm/2.75) was defloyed at the middle-to-distal LAD lesion.

After stent insertion, resolution of the intimal dissection area was confirmed by IVUS and coronary angiography (Fig. 1B, Fig. 2B).

DISCUSSION

SCAD is a rare, but very dangerous disease of the coronary arterial wall that occurs without any major cardiovascular risk factors. SCAD reported in only 0.07-1.10% of patients referred for coronary angiography. The first case of SCAD was reported in 1931, while first the angiography-confirmed case was reported in 1978.[1] SCAD mainly occurs in younger women, and atherosclerosis, fibromuscular dysplasia, pregnancy, and the postpartum period are the known risk factors. In addition, changes to the arterial wall due to hormonal influences during pregnancy are thought to be involved.[6] Other rare causes include connective tissue disorders (Marfan syndrome, Loeys-Dietz syndrome, Ehlers-Danlos syndrome, systemic lupus erythematous and so on.) and systemic inflammatory

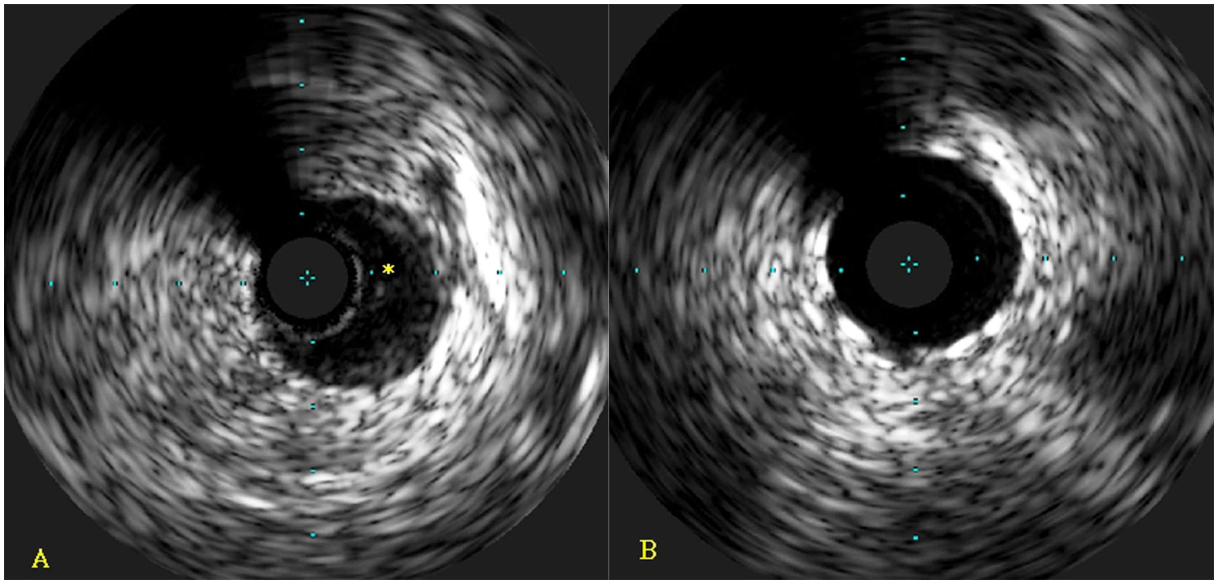

Fig. 2A : Intimal tear and hematoma(*) of the left anterior descending artery, revealed by intravascular ultrasonography
2B : Resolution of the intimal dissection after stent insertion

conditions, hormonal therapy, drug use, and physical stress.[2]

SCAD is mainly diagnosed by coronary angiography. In some cases, optical coherence tomography, or IVUS can be used for diagnosis. Non-invasive methods such as computed tomography or magnetic resonance angiography can miss small or distal arteries.

FXS is the most common genetic disorder that causes mental retardation. FXS is caused by the hyperexpansion of a polymorphic CGG trinucleotide repeat in the 5' untranslated region of the fragile X mental retardation-1 (FMR1) gene. FXS patients are known to have various medical complications such as otitis media, sinusitis, mitral valve prolapse, seizure, and so on. Moreover, women with FXS permutation reportedly have an increased risk of premature ovarian failure (21-33%).[7]

In the present case, besides the FXS permutation, the patient did not have any other underlying conditions. The patient experienced early menopause 5 years prior, and underwent several years of hormonal therapy. There are no reports of a direct relationship between FXS and SCAD. However, early menopause and hormonal therapy could play a role in SCAD.

Medical treatment is considered to be standard for SCAD when there is no change in the electrocardiogram or the patient is stable. Medical options for the treatment of SCAD include aspirin and beta-blockers. Unstable patients or patients exhibiting symptoms may benefit from stent insertion or bypass surgery.[8]

In our patient, ST segment elevation, and chest pain symptom was observed to continue, Coronary angiography revealed that distal flow is bad, so was PCI. In a recent report, SCAD patients underwent PCI of getting success rate was reported as

30%.[8] However, treatment results when using the DES have not been reported. It has recently been introduced a bioabsorbable coronary stents. Therefore, it will require a multicenter study to assess the PCI treatment results in patients with SCAD.

SCAD is a rare, but important cause of sudden cardiac death in younger women. The condition is associated with a number of risk factor, such as connective tissue diseases, vasculitis, inflammatory disorders, congenital disorders and physical stress. In addition, fibromuscular dysplasia (FMD) also have been reported as rare causes of SCAD.[9]

In our institution, we have encountered certain patients with FXS. The relationship between FXS and SCAD is unclear. However, women with no evident coronary risk factors may develop SCAD. Therefore, A variety of causes (FMD, early menopause, connective tissue disease, and so on.) associated with FXS needs to be explored further.

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