CASE REPORT

Spontaneous multivessel coronary artery dissection with anomalous coronary artery

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SUMMARY

Spontaneous coronary artery dissection (SCAD) is one of the rare causes of acute coronary syndrome in young healthy individuals especially women without having any conventional risk factors for coronary artery disease. We describe a case of 34-year-old healthy man with diffuse multiple SCADs who presented with acute coronary syndrome and was managed conservatively with an uneventful course on long-term follow-up.

BACKGROUND

Spontaneous coronary artery dissection (SCAD), a rare cause of acute coronary syndrome, was first described in 1931 in a 42-year-old white woman who had sudden cardiac death following chest pain.¹ Even though the single coronary artery involvement is usually seen in SCAD, the left main coronary artery (LMCA) with multiple coronary involvement is still a rare finding. We describe a case of SCAD involving multiple coronaries with an associated anomalous coronary artery which was managed conservatively. We highlight the rarity of diffuse involvement of multiple coronary arteries with an associated anomalous coronary artery and the description of various management issues.

CASE PRESENTATION

A 34-year-old man was admitted to a peripheral hospital with acute anterior wall myocardial infarction and was thrombolysed with streptokinase. Subsequently, he was referred to our hospital for further management. He was a healthy, non-smoker without any history of illicit drug use. No significant family history was noted.

INVESTIGATIONS

ECG showed normal sinus rhythm with the QS-pattern in precordial leads V1–V5. Transthoracic echocardiogram revealed regional wall motion abnormalities in left anterior descending (LAD) artery territory with mild global left ventricular dysfunction (ejection fraction of 45%). The patient was taken up for coronary angiogram after informed consent. Selective left coronary angiogram revealed multiple dissections involving the whole length of the LMCA, LAD and major diagonal up to their mid-segment with distal TIMI-III (thrombolysis in myocardial infarction) flow (figure 1A and B; see online video 1). Selective right coronary sinus shoot (figure 1C; see online video 2) revealed normal right coronary artery (RCA), anomalous origin of left circumflex (LCX) from right sinus with dissection in its mid-segment.

Arch aortogram revealed normal arch vessels. Renal angiogram was normal.

DIFFERENTIAL DIAGNOSIS

Primary non-traumatic SCAD is one of the rare causes of acute coronary syndrome or sudden cardiac death in the young otherwise healthy individual with a special predilection for women. Fibromuscular dysplasia (FMD), a segmental non-atherosclerotic non-inflammatory angiopathy of unknown aetiology, is a novel association and potentially causative factor for SCAD.^{2–3} Recently, a study has observed that the FMD is an important risk factor for SCAD and that the true prevalence of FMD was likely to be higher than identified.⁴ FMD is seen in young women, commonly involving renal and carotid arteries. Coronary FMD commonly is overlooked, as the angiographic appearance is often subtle. Other possible associations with SCAD are connective tissue disorders namely Marfan's syndrome and Ehlers-Danlos type IV, systemic lupus erythematosus, etc. But, our patient did not have any peripheral stigmata suggestive of these systemic disorders.

TREATMENT

Our patient was asymptomatic and haemodynamically stable. So, in view of the extensive nature of dissection involving multiple coronary arteries, but with adequate TIMI flow, we planned for the conservative management. The patient was kept under observation in the hospital for 10 days and was discharged on dual antiplatelet drugs, β blocker, ACE inhibitor and statin.

OUTCOME AND FOLLOW-UP

Stress myocardial perfusion imaging done after 1 month showed no evidence of viable myocardium in LAD territory. After 9 months, the coronary check angiogram revealed persistence of similar extent of dissection involving same coronary arterial segments as previous (figure 2A and B; see online video 3). But, as the patient was asymptomatic, we decided to continue only with conservative management. At 24-months of follow-up, the patient is doing well without any complication.

DISCUSSION

Primary non-traumatic SCAD is one of the rare causes of acute coronary syndrome or sudden cardiac death in the young otherwise healthy individual with a special predilection for women. The majority (80%) occurs in young women during the peripartum period or in association with oral

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Figure 1 Selective left coronary angiogram in the right anterior oblique caudal (A) and cranial (B) projections shows multiple spontaneous dissections involving the whole length of the left main coronary artery (thick white arrows), left anterior descending artery upto mid-segment (thin white arrows) and major diagonal (black arrows) upto its mid-segment with distal TIMI (thrombolysis in myocardial infarction) III flow. Selective right coronary angiogram (C) shows anomalous origin of left circumflex from the right coronary sinus with evidence of dissection just before the first obtuse marginal branch (black arrow). Right coronary artery is normal. LMCA, left main coronary artery; LAD, left anterior descending; D1, first diagonal; RCA, right coronary artery; LCX, left circumflex.

contraceptive use, without any conventional risk factors for coronary artery disease (CAD) or atherosclerosis. Men frequently present at a slightly later age and usually have evidence of CAD or risk factors for atherosclerosis.⁵ Prognosis is somewhat better for men as compared to women. In general, the long-term prognosis of patients with SCAD is favourable if they survive the acute phase,^{6, 7} as observed in the case of our patient. In a review comprising 152 cases over the last 50-years, Kamineni *et al*⁸ reported that 50% of patients with SCAD developed recurrence within 2 months. Overall, LAD artery is affected in 75%, RCA in 20%, LCX artery in about 4% and the LMCA in <1% of the cases.⁹ Approximately 70% of these patients have dissection in a single coronary artery only. Spontaneous dissection of multiple coronary vessels with LMCA involvement, as in the case of our patient, is very rare. Various diagnostic modalities can be employed to diagnose SCAD. CT angiography is a non-invasive modality which could be used for the follow-up as well. Apart from that, conventional coronary angiogram can also be used as we have used in our patient's case. Intravascular ultrasound (IVUS), though invasive, has recently been considered as an essential mode of investigation for both the diagnosis and follow-up. Furthermore, IVUS helps to identify the signs of coronary atherosclerosis. Although the medical management

and revascularization, either percutaneously or surgically, are available treatment modalities in these patients, the optimal strategy for this disease process has not been clearly defined.¹⁰ The choice of the appropriate therapy is primarily based on the clinical presentation, the extent of the dissection and the amount of myocardium at risk.¹¹ Thrombolytic agents as first-line therapy are avoided as they may increase the extent of dissection. A high index of suspicion should be kept to consider SCAD as the probable cause of acute coronary syndrome, especially in those subset of patients who are not having the conventional risk factors for CAD. Surgical intervention is indicated in those cases that demonstrate evolving dissections and significant haemodynamic compromise.¹¹ Surgical limitations include difficulty in finding the true lumen and associated non-negligible mortality. On the contrary, percutaneous coronary intervention is highly operator dependent and is associated with complications such as difficulties while advancing the guidewire through the true lumen, proximal and distal progression of the dissection, and acute closure of the vessel. IVUS can be very useful during percutaneous intervention in ascertaining the proper position of the guidewire in the true lumen. Patients with non-occlusive coronary dissections with the normal antegrade coronary flow, without any haemodynamic instability or continuing

Video 1 Selective left coronary angiogram in right anterior oblique caudal projections showing multiple spontaneous dissections.

Video 2 Selective right coronary angiogram showing the anomalous origin of left circumflex from the right coronary sinus with evidence of dissection just before the first obtuse marginal branch.

Figure 2 At nine month follow-up, coronary angiogram in the right anterior oblique caudal (A) and cranial (B) projections showed multiple dissections involving whole length of LMCA (thick white arrows), LAD artery upto mid-segment (thin white arrows) and D1 (black arrows) upto its mid-segment with distal TIMI III flow. LMCA, left main coronary artery; LAD, left anterior descending; D1, first diagonal.

symptoms or evidence of viable myocardium, should be treated with conservative management, as most of these patients have shown complete healing of the dissection on follow-up angiography at a later stage. Medical therapy did not differ from standard therapy for patients with acute coronary syndromes, that is, dual antiplatelets, anticoagulation either with unfractionated heparin or low-molecular-weight heparin.

Learning points

- ▶ Spontaneous coronary artery dissection (SCAD) is one of the rare cause of acute coronary syndrome in young healthy individuals, especially women without having any conventional risk factors for coronary artery disease.
- ▶ In SCAD, left anterior descending artery is the most common (75%) artery involved followed by right coronary artery (20%), left circumflex artery (4%) and left main coronary artery (<1%).
- ▶ Fibromuscular dysplasia, a segmental non-atherosclerotic non-inflammatory angiopathy of unknown aetiology, is a novel association and potentially causative factor for SCAD.
- ▶ The natural history of SCAD includes spontaneous healing with complete resolution.

Video 3 At nine month follow up, coronary angiogram showing persistent coronary dissection.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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