

Available online at [www.sciencedirect.com](http://www.sciencedirect.com)**ScienceDirect**journal homepage: [www.elsevier.com/locate/ihj](http://www.elsevier.com/locate/ihj)**Original Article****Spontaneous coronary artery dissection: Case series and review of literature**Satyavan Sharma<sup>a</sup>, Nikhil Raut<sup>b,\*</sup>, Anil Potdar<sup>c</sup><sup>a</sup> Professor and Head of Cardiology and Interventional Cardiologist, Department of Cardiology, Bombay Hospital and Bombay Hospital Institute of Medical Sciences, Mumbai, India<sup>b</sup> Senior Registrar, Department of Cardiology, Bombay Hospital and Bombay Hospital Institute of Medical Sciences, Mumbai, India<sup>c</sup> Honorary Cardiologist, Department of Cardiology, Bombay Hospital and Bombay Hospital Institute of Medical Sciences, Mumbai, India**ARTICLE INFO****Article history:**

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**ABSTRACT**

**Aim:** Spontaneous coronary artery dissection (SCAD) is a less recognized cause of ST elevation myocardial infarction (STEMI) in clinical practice. The aim of this communication is to describe a case series in South Asian population and highlight on the long-term clinical outcomes on conservative management.

**Methods:** A retrospective analysis of data of five patients (6 instances) of SCAD managed in a tertiary care center during January 1994 to June 2015 was done. Clinical, angiographic, therapeutic, and follow-up data till end of June 2015 are analyzed.

**Results:** All patients were young (mean – 33 years) and predominantly male. Etiology of SCAD was diverse and included peripartum state, vigorous activity and atherosclerosis. Left anterior descending (LAD) coronary artery was predominantly involved and the majority had angiographic type 1 SCAD. Medical treatment provides excellent long-term benefits. Coronary stenting provided symptomatic benefit in a patient with favorable anatomy.

**Conclusions:** Clinical recognition of SCAD is difficult. It should be suspected in peripartum state, young females and in presence of other precipitating factors. Coronary angiography is essential for establishing the diagnosis. Medical treatment provides favorable long-term survival.

**Implications and practice:** The awareness of SCAD is important for all clinicians involved in STEMI care. A prompt suspicion can avoid administration of thrombolytic therapy. Early coronary angiography will provide an accurate diagnosis and help in deciding appropriate therapy. Percutaneous intervention can be challenging.

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## 1. Introduction

Spontaneous coronary artery dissection (SCAD) is a rare clinical entity that frequently presents as an acute coronary syndrome (ACS).<sup>1,2</sup> Dissection of the coronary artery results in separation of the layers of the arterial wall, creating a false lumen. The separation may be between the intima and media, or between the media and adventitia. Hemorrhage into the false lumen can impinge upon the true lumen of the coronary artery, impairing the blood flow and causing myocardial ischemia, infarction, sudden death, cardiogenic shock, or pericardial tamponade.<sup>1,3,4</sup>

SCAD is often classified according to the associated predisposing condition and has been broadly divided into atherosclerotic and nonatherosclerotic. Nonatherosclerotic associations have included peripartum state, connective tissue disorders, systemic inflammatory conditions, and coronary artery spasm.<sup>5,6</sup> Fibromuscular dysplasia (FMD) as a possible predisposing factor particularly in females has received a lot of attention recently.<sup>5-9</sup> Screening for FMD of renal, iliac, and cerebrovascular arteries by computed tomography angiography and magnetic resonance angiography can be helpful. Association of extracoronary vascular abnormalities like dissections, aneurysms, and dilatation in chest, abdomen, pelvis, and neck has also been described.<sup>7</sup> Those without identifiable predisposition have been labeled as idiopathic. Vigorous exercise with increased cardio-circulatory stress and shear forces against the coronary arterial wall can precipitate SCAD among patients with or without these predisposing conditions.<sup>3,4</sup>

Diagnosis of SCAD is made at coronary angiography.<sup>3,4,6,8,9</sup> Recent reports suggest the value of intravascular ultrasound (IVUS) and optical coherence tomography (OCT) for diagnosis and management.<sup>6,8-12</sup> Various treatment options are available including medical therapy, percutaneous coronary intervention (PCI), and coronary artery bypass graft (CABG) surgery.<sup>3,4,6</sup>

There are isolated case reports on this entity from South Asia.<sup>12,13</sup> This communication reports a series of 5 cases from South Asia with diagnostic, therapeutic, and follow-up data along with brief review of the literature.

## 2. Methods

The material for this study is obtained from records of academically interesting patients maintained by the first

author in a tertiary care referral postgraduate teaching institute. During January 1994 to June 2015, five cases of SCAD were diagnosed, treated, and followed up. The records of these cases form the material for this analysis.

The diagnosis of SCAD was made on the basis of the following angiographic features<sup>1,3,4,6,8,9</sup>:

- (1) Visualization of thin radiolucent intimal flap and the presence of extraluminal contrast after wash out of dye from the remainder of the vessel.
- (2) Presence of haziness and irregularity of the vessel. Haziness and intraluminal filling defects indicate possibility of a thrombus.

Patients were classified into types 1, 2, and 3, as per the angiographic classification proposed by Yip<sup>6</sup> and Saw.<sup>14</sup> Type 1 is pathognomic angiographic appearance of SCAD with contrast dye staining of arterial wall with multiple radiolucent lumen. Type 2 includes lesions with long, diffuse, and smooth stenosis representing intramural hematoma. Type 3 mimics atherosclerosis.

History of pregnancy, vigorous exercise, connective tissue disorder, substance abuse, diabetes and hypertension was noted. Clinical data, electrocardiogram (ECG), two-dimensional echocardiography (2DE), management details, and follow-up information were analyzed. In four patients, full blood analysis, including blood count, total body profile, coagulation workup, lipoprotein assay, thyroid function test, and a workup for autoimmune disorder, was available. One patient had repeat coronary angiography at one year and a multidetector computed tomography (MDCT) coronary angiography at 2 years follow-up.

Patients with coronary dissection who had any history of cardiac surgery, coronary intervention, trauma, or aortic dissection were excluded.

## 3. Results

The patient characteristics are summarized in [Table 1](#). The patients were from different states of India. Cases 1 and 5 were from Maharashtra, whereas cases 2 and 3 were referred from Madhya Pradesh and case 4 from Rajasthan. Four patients were male and ages ranged from 18 to 56 years, mean – 33 years. There were 6 instances of SCAD in 5 patients. All patients presented with ST elevation on ECG. Case 1 was treated during both the episodes at our center whereas the

**Table 1 – Clinical data.**

| Case number    | Year       | Age (years) | Sex | Presentation       | Follow-up (years) | Predisposing/Precipitating factor |
|----------------|------------|-------------|-----|--------------------|-------------------|-----------------------------------|
| 1 <sup>a</sup> | 1994; 2011 | 39; 56      | F   | AW-STEMI; IW-STEMI | 19                | Pregnancy; DM, HTN                |
| 2              | 1998       | 18          | M   | AW-STEMI           | 5                 | No obvious cause                  |
| 3              | 1998       | 19          | M   | AW-STEMI           | 5                 | Vigorous activity                 |
| 4              | 2011       | 40          | M   | IW-STEMI           | 4                 | DM, HTN, ↑homocysteine            |
| 5              | 2013       | 31          | M   | AW-STEMI           | 2                 | Tobacco, ↑homocysteine            |

Abbreviations: AW, anterior wall; DM, diabetes mellitus; F, Female; HTN, hypertension; IW, Inferior wall; M, Male; STEMI, ST elevation myocardial infarction.

<sup>a</sup> Case 1 had an anterior wall STEMI in 1994 and inferior wall in 2011.

**Table 2 – Angiographic data and therapy.**

| Case number    | Vessel     | Segment involved             | Type of SCAD | Therapy       |
|----------------|------------|------------------------------|--------------|---------------|
| 1 <sup>a</sup> | LAD; (RCA) | Proximal; (Proximal and mid) | 1            | Medical       |
| 2              | LAD        | Distal                       | 2            | Medical       |
| 3              | LAD        | Proximal and mid             | 1            | Medical       |
| 4              | RCA        | Proximal, mid and distal     | 1            | Medical       |
| 5              | LAD        | Proximal and mid             | 1            | Medical + PCI |

Abbreviations as in text.  
<sup>a</sup> Case 1 had LAD involvement during first MI and RCA during the second episode.

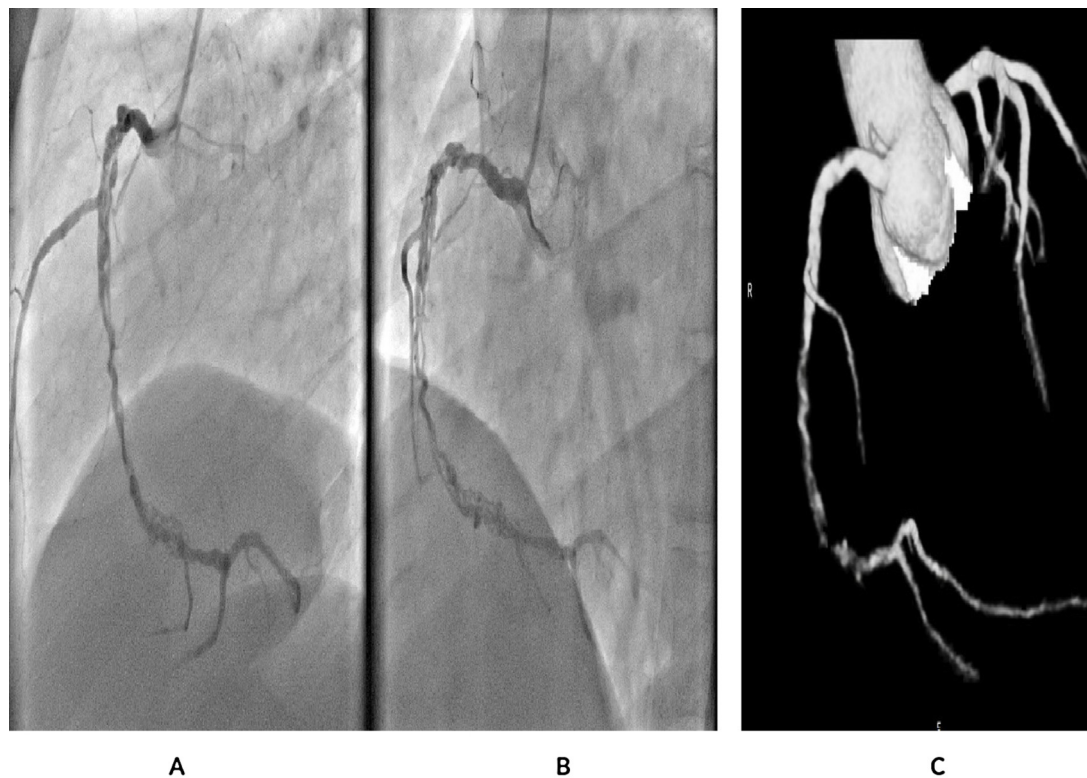
remaining patients were referred for coronary angiography after initial treatment.

Case 1 had anterior wall ST elevation myocardial infarction (STEMI) at the age of 39 years, fifteen days after an uneventful delivery and an inferior wall STEMI at the age of 56 years. Cases 2, 4, and 5 had received thrombolysis with streptokinase in an outside hospital. The predisposing factor for SCAD was peripartum state in case 1 and possible precipitation in case 3 by vigorous activity (not trauma) while playing cricket. Complete blood analysis as detailed in methods was normal in all cases except for elevated homocysteine in cases 4 and 5. Cardiovascular risk factors like diabetes and hypertension (case 1 during second episode and case 4) and tobacco consumption (case 5) were observed in 3 patients. The mean left ventricular ejection fraction on 2DE was 46% (30–50%).

Angiographic data are summarized in Table 2. The left anterior descending (LAD) artery was involved in all instances

of anterior wall and right coronary artery (RCA) in inferior wall STEMI. Involvement of proximal and mid segment was seen in 5 instances. Majority had type 1 SCAD. All patients received optimal medical therapy using antiplatelet, statins, beta-blocker, angiotensin-converting enzyme inhibitors, and other drugs. Case 5 had PCI using 3 mm × 28 mm paclitaxel eluting (Taxus Liberte) stent (Fig. 2).

The mean follow-up duration was 7 (range 2–19) years (Table 1). Case 1 had recurrence of STEMI in another territory after 17 years. On angiography, she had complete resolution of dissection in the LAD and had SCAD in RCA. Cases 2 and 3 did extremely well clinically for 5 years and were subsequently lost to follow-up. Case 4 with extensive SCAD in entire RCA remains asymptomatic. Follow-up coronary angiography at 1 year (Fig. 1B) and MDCT coronary angiography (Fig. 1C) at 2 years revealed no change in angiographic characteristics. Case 5 is asymptomatic 2 years after LAD stenting.



**Fig. 1 – RCA angiography in left anterior oblique (LAO) cranial view documents extensive type 1 SCAD of RCA from proximal to distal segment in case 4 at presentation (panel A), at 1 year follow-up (panel B). CT coronary angiography (panel C) at 2 years shows no resolution of the dissection. Abbreviations as in text.**

#### 4. Discussion

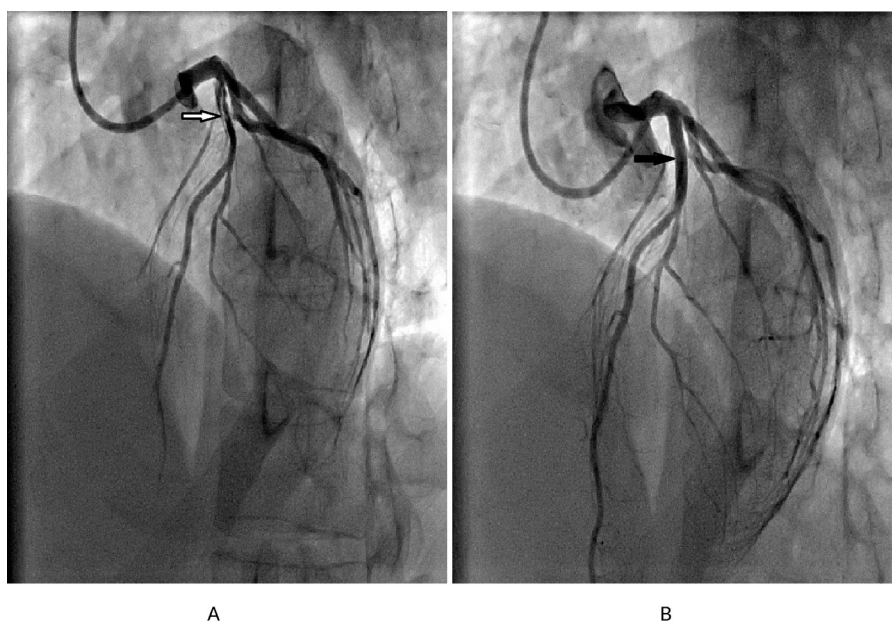
SCAD has been well documented in several studies in Caucasian patients.<sup>1-6,8-11,14-18</sup> There is paucity of data in South Asian population and this communication describes a case series of SCAD from this subcontinent.

Five young patients with STEMI who presented to a tertiary care center for management were found to have SCAD on coronary angiography. Clinical diagnosis of SCAD is difficult and 70% of the reported cases are women.<sup>4</sup> Diagnosis of SCAD should be strongly considered in a woman who has chest pain or ACS during pregnancy. In this study, only one patient was female (case 1) and she had SCAD twice. The first event was an anterior wall STEMI 2 weeks after the delivery. One-third of all SCAD cases in women occur in the peripartum period, of which one-third occur in late pregnancy and two-thirds in early postpartum period.<sup>4,13</sup> The pathogenesis of SCAD in peripartum period is unclear. Hemodynamic factors together with arterial wall changes related to alterations in sex hormones predispose to intimal tear and SCAD. The etiology of second episode of SCAD in right coronary artery could have been atherosclerosis, connective tissue disorder, or FMD. There was no history of connective tissue disorder. Many recent studies have suggested FMD as a possible predisposing factor for SCAD in young females.<sup>5-9</sup> FMD is a nonatherosclerotic and noninflammatory disease involving renal, carotid, iliac, and coronary vessels. The most common angiographic appearance in FMD is "string of beads" appearance due to medial hyperplasia. The second STEMI in this case is unlikely to be due to FMD, although the same cannot be ruled out in absence of systematic screening of renal, iliac, and cerebrovascular vessels. Atherosclerosis as a possible cause is strengthened by age of the patient and presence of atherosclerotic risk factors diabetes and hypertension.

Interestingly, majority of the patients in this study were male. There was no history of connective tissue disorder or cocaine use. All these patients were diagnosed as SCAD during the years 1994–2013 when awareness about FMD as a possible cause was limited. The mechanism of SCAD in these patients remains unclear and diverse. It is likely that vigorous exercise precipitated SCAD in case 3, who developed STEMI 2 h after a cricket match. The SCAD in cases 4 and 5 was probably atherosclerotic, as these patients had cardiovascular risk factors and premature atherosclerosis is common in our country. Atherosclerotic plaque inflammation and rupture may cause disruption of intimal-medial junction resulting in intimal flap and subsequent hematoma formation. No obvious factor could be identified as cause of SCAD in case 2.

The diagnosis of SCAD on coronary angiography is challenging.<sup>1,3,4,6,8,9</sup> The possibility should be considered in a young woman with ACS or circumstances predisposing to SCAD. There are distinctive features that facilitate prompt diagnosis and appearances where recognition may be difficult. The typical angiographic appearance is of thin longitudinal radiolucent line representing an intimal medial flap with flow in two or more separate lumens (Figs. 1 and 2). The intramural lumen may fill and empty slowly and may distally end in a cul-de-sac with stasis of dye in between the injections. The intraluminal dissection flap may show haziness and filling defects suggestive of thrombi. SCAD has been classified into three types<sup>6,14</sup> depending on angiographic appearance. Type 1 is easily diagnosed (Figs. 1 and 2). Long diffuse and long smooth stenosis representing intramural hematoma (type 2) can be diagnosed by experienced operators. However, cases with shorter (20–30 mm) lengths of SCAD and type 3 SCAD often require intracoronary imaging for confirmation of diagnosis.

The LAD involvement was seen in 4 out of 6 instances (66%). In angiographic series, the LAD accounts for average 60% (38–77%) of cases.<sup>4</sup> Several cases of SCAD with multivessel



**Fig. 2 – Left coronary angiography in left anterior oblique with cranial view shows type 1 SCAD of LAD (white arrow) in case 5 (panel A). Panel B shows sealing of dissection and excellent flow after coronary stenting (black arrow) (panel B). Abbreviations as in text.**



and left main coronary artery (LMCA) involvement have been reported. Cannulation of the coronary ostium should be done with great caution in patients suspected of having a SCAD, as forceful contrast injections during coronary angiography may lead to extension of the dissection.<sup>4</sup>

There is limited use of MDCT in the diagnosis of SCAD due to lower spatial resolution compared to conventional angiography.<sup>15</sup> The best use of this modality is for follow-up of patients in whom a conservative management is chosen as in case 4.

Coronary angiography is extremely valuable in diagnosis but fails to provide details of the coronary wall. Recent reports suggest the value of IVUS and OCT in the diagnosis and management.<sup>6,8–12</sup> Typical IVUS features are the presence of intramural hematoma in the outer one-third of the media compressing the true lumen with minimal or no atherosclerosis. The very high-resolution images obtained on OCT provide unique insights into intimal tear, intimo-medial flap, double lumen morphology, intramural hematoma, and associated thrombus. The technique can also guide coronary intervention. OCT use in this unstable anatomic subset may be risky and the use should be restricted to expert operators.

The understanding of SCAD has improved in recent years and it is important to incorporate this into our future practice. Due to strong association of FMD with SCAD in many studies,<sup>5–9</sup> all patients with SCAD should undergo a systematic screening for FMD in iliac, renal, and cerebral vessels using appropriate imaging modality.

There are no specific guidelines on how to manage patients with SCAD. Treatment options for SCAD include medical therapy, PCI, or CABG surgery.<sup>3,4,6</sup> Medical management of SCAD is similar to the treatment of ACS. It includes antithrombotic therapy with heparin or low-molecular weight heparin, aspirin, clopidogrel, glycoprotein IIb/IIIa inhibitors, and anti-ischemic therapy. However, the use of a potent antithrombotic therapy in SCAD may be a double-edged sword; on one hand it will help to decrease thrombus formation in the false lumen and thereby improve blood flow in the true lumen, on the other hand, it may increase bleeding in the false lumen, causing an expansion of intramural hematoma, resulting in a decreased flow through the true lumen. Fibrinolysis is not recommended due to increased bleeding and advancing dissection.<sup>4,16</sup> Unfortunately, the converse occurs in day-to-day practice due to difficulties in clinical diagnosis of SCAD. Three patients in this series had received thrombolysis. Thrombolysis should be avoided in patients predisposed to SCAD as in cases 1 and 3.

A conservative approach is generally favored in stable SCAD patients, as was in this series. The conservative strategy can provide excellent long-term follow-up.<sup>6,8,9</sup> During follow-up (mean – 7 years), no patient had sudden cardiac death. One patient had STEMI in another territory after 17 years. With conservative management, partial or even complete angiographic resolution of dissections has been observed during follow-up.<sup>6,8,9,17</sup>

The decision to revascularize with PCI or CABG should depend on clinical status, hemodynamic instability, and angiographic characteristics. In this series, PCI was performed in one case with ongoing symptoms and favorable anatomy. Technical difficulties during PCI include advancing the guide-wire in the true lumen rather than the false lumen, distal

propagation of the intramural hematoma, and dissection during stent delivery.<sup>6,8,9</sup> Mayo case series<sup>18</sup> documented a technical success rate of only 65% with PCI; furthermore, 25% of successful PCI had propagation of the dissection requiring additional stents. OCT or IVUS can guide wire placement in the true lumen, evaluate the length of dissection, vessel size, and confirm stent apposition and dissection sealing. At present, routine use of PCI is not recommended. Surgical revascularization is indicated with multivessel or LMCA involvement or refractory ischemia. Graft placement can be challenging in cases of SCAD involving a long coronary artery segment.<sup>9</sup>

## 5. Conclusions

This is a first case series with long-term follow-up data documenting SCAD as a cause of STEMI in South Asian population. Etiology of SCAD is multifactorial. Most patients in this study were male in contrast to western literature. Early coronary angiography can facilitate prompt recognition and avoid thrombolysis. The long-term prognosis is favorable on conservative treatment. This is a selective case series and has inherent limitations. The study does not provide any information about prevalence of SCAD in ACS or in patients undergoing coronary angiography.

## Conflicts of interest

The authors have none to declare.

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