

Spontaneous coronary artery dissection

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A 38 year old previously healthy woman was referred to ER for ongoing chest pain. She had sudden onset of central, crushing chest pain 3 h back and her ECG was found to have evidence of acute infero-lateral wall MI. There was no history of cardiovascular disease or identifiable CAD risk factors. She had a history of bone TB 10 y back. She was on oral contraceptive pills 5 y back. Her mother had hypertension and diabetes mellitus. Her father died at the age of 75 y due to unknown cause and had a history of hypertension. There is no history of hypertension, diabetes mellitus or CVD in her siblings.

General Examination: O₂ saturation-95%, Pulse-88bpm, Bp-110/90mmHg. Systemic Examination: no abnormality detected. Investigation: CBC, RFT, BSR, and Electrolytes were WNL. ECG: ST elevation in infero-lateral leads 3 h back but minimal ST elevation in III and AVF in our ER. Cardiac Biomarkers: CPK-MB-50u/l and Trop +. Echo screening: hypokinetic apex, apical IVS and apical inferior LV wall.

She was diagnosed as acute infero-lateral wall MI and was admitted in CCU was treated with Aspirin, Clopidogrel, LMWH, Atorvastatin, IV GTN, Beta-Blocker, Anxiolytics, PPI and Stool softener. She was well on first post MI day but suddenly developed chest pain on second post MI day and her ECG showed ST elevation in V3-V6. She was taken to Cath lab for rescue PCI. Her coronary angiography studies revealed a linear dissection involving the distal LAD with TIMI 3 distal flow (Fig1). She was conservatively management and was discharged on 7 post MI day.

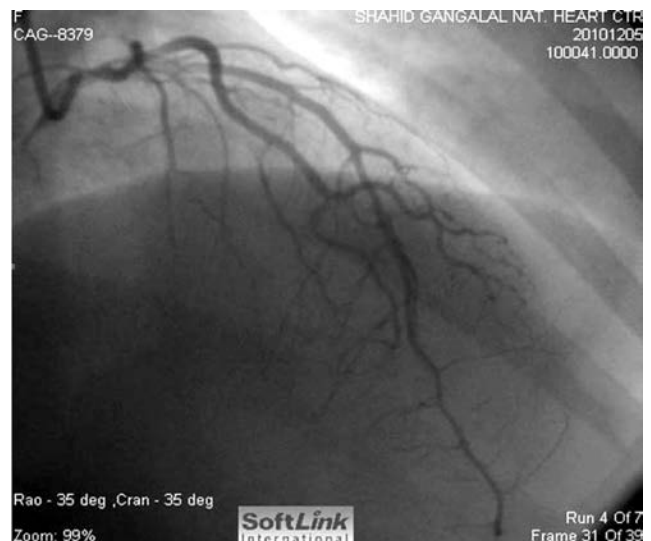
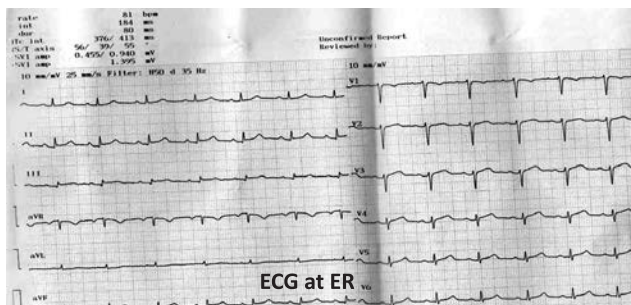
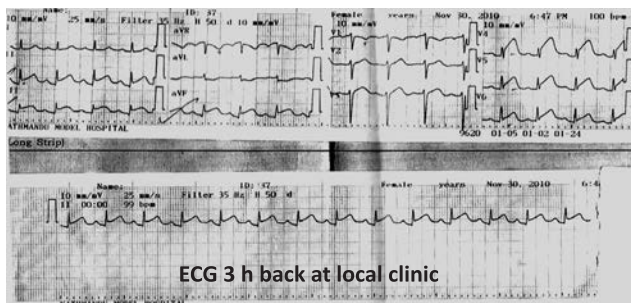


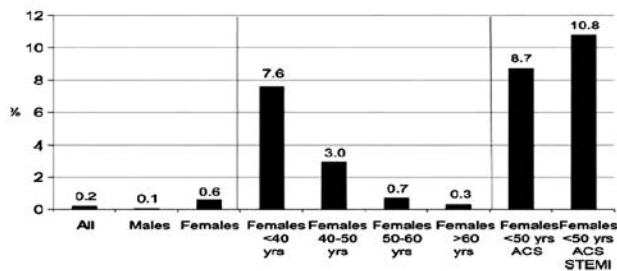
Fig 1. Dissection in the distal LAD with TIMI 3 distal flow.

Spontaneous coronary artery dissection

Spontaneous coronary artery dissection (SCAD) is a very rare cause of acute coronary syndromes (1). SCAD can be primary or secondary. Primary dissections occur spontaneously whereas secondary dissections occur as an extension from aortic root dissection or following an insult as a consequence of coronary angiography, coronary intervention, cardiac surgery or chest trauma(2).

Incidence

The first case of SCAD was described in 1931 by Pretty in a 42 year old woman who had a sudden cardiac death (3). Initial cases were based on autopsy findings after sudden cardiac death. The incidence of SCAD in angiographic series



varies widely from 0.07% up to 1.1% for patients who are referred for coronary angiography (4). The mean age at presentation is 30-45 years (5). More than 70% of SCAD cases are women, and in approximately 30% it occurs during the peripartum period (6). Among women, the incidence of SCAD was highest in women below the age of 40 years (Fig 2) and decreased significantly with advancing age (4). Although not invariable, left coronary artery dissections are more common in women, the RCA is usually involved in men. Overall, the LAD artery is affected in 75% of cases, the RCA in 20% of cases, the left circumflex artery in about 4% of cases, the LMCA in < 1% of cases and multivessel coronary dissection was observed in about 20% (7).

Pathogenesis and etiology

SCAD remains an unclear aetiopathologic entity. SCAD results from vessel wall haematoma formation in the outer third of the media or between the media and the adventitia in the absence of traumatic or iatrogenic causes, resulting in a false lumen (8). Expansion of this lumen through blood or clot accumulation leads to distal propagation of the dissection and to compression of the real lumen, causing myocardial ischemia (9). An intimal tear is only seldom observed.

The most common pathologies associated with SCAD are coronary atherosclerosis and vascular changes occurring during the peripartum period. Other causes of SCAD are

connective tissue disorders, systemic lupus erythematosus vasculitis, cocaine abuse, vigorous exercise, and prolonged sneezing. However, a large number of cases must be classified as idiopathic because no underlying condition can be detected.

SCAD in the peripartum period

The pathogenesis of SCAD in the peripartum period is still unclear. Hemodynamic factors together with arterial wall changes related to pregnancy, a lytic action of proteases released from eosinophils, and intimal tears are the main hypotheses presented to explain the pathophysiology involved (10). Changes in the concentrations of sex hormones are thought to alter the normal arterial wall architecture, resulting in an increased susceptibility to spontaneous dissections (11). The changes in the vascular wall include smooth muscle cell proliferation, impaired collagen synthesis, and alterations in the protein and acid mucopolysaccharide content of the media.

During pregnancy total blood volume and cardiac output are increased. This may lead to augmented shear forces on the luminal surface and an increased wall stress in pregnancy and particularly during labour. Both vascular and hemodynamic changes occurring during pregnancy and labour therefore predispose the coronary arteries to the development of intramural dissections (12).

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 the early postpartum period (11). The peak incidence is within the first 2 weeks after delivery. The earliest reported case presented at 9 weeks of conception and the latest 3 months postpartum. Only 30% of the patients in this group have known risk factors for coronary artery disease (11). Patients with advancing age and multiparity have an increased risk for SCAD (11).

In a population based study of more than 12 million deliveries in the USA, the incidence of acute myocardial infarction was 6.2 per 100 000; 45% of the women diagnosed with acute myocardial infarction underwent coronary angiography and 37% have undergone a revascularization procedure. A recent review of the literature revealed a high incidence of risk factors for ischaemic heart disease in patients with pregnancy associated myocardial infarction. Evaluation of coronary artery morphology (angiographically or at autopsy) revealed a dissection only in 28% of the patients, whereas a coronary stenosis was observed in 40%. Coronary dissection was the primary cause of infarction in the peripartum period (50%) and was found more commonly in postpartum compared with antepartum cases 34% vs 11%(13).

Clinical Presentation

The clinical presentation ranges from unstable angina, acute myocardial infarction, ventricular arrhythmias to sudden cardiac death (14). In rare instances it can be asymptomatic and discovered incidentally on coronary angiography. Whenever a young patient without major coronary risk factors or a woman in the postpartum period presents with an acute coronary syndrome or sudden cardiac death, the possibility of a SCAD should be suspected and an urgent coronary angiography considered.

Diagnosis

Early coronary angiography remains essential in the diagnosis of SCAD (15). Additional intravascular ultrasound (IVUS) and Optical coherence tomography (OCT) imaging provides very detailed information on the location and extent of the dissection (16, 17). CT coronary angiography is useful in the follow-up of conservatively managed patients.

Coronary angiography

Angiographically coronary dissections can be graded according to the National Heart, Lung, and Blood Institute classification system developed by the Coronary Angioplasty Registry (15). This system grades coronary dissections based on angiographic appearance as types A-F. Type A dissections represent radiolucent areas within the coronary lumen during contrast injection, with minimal or no persistence of contrast after the dye has cleared. Type B dissections are parallel tracts or double lumen separated by a radiolucent area during contrast injection, with minimal or no persistence after dye clearance. Type C dissections appear angiographically as contrast outside the coronary lumen, with persistence of contrast in the area after clearance of dye from the coronary lumen. Type D dissections represent spiral luminal filling defects, frequently with extensive contrast staining of the vessel. Type E dissections appear as new, persistent filling defects. Type F dissections represent those that lead to total occlusion of the coronary artery, without distal antegrade flow.

Management

There is no specific guideline on how to manage patients with SCAD. Treatment options for SCAD include medical therapy, percutaneous coronary intervention (PCI), or coronary artery bypass graft surgery (CABG). The decision to manage SCAD conservatively with medication or to perform PCI or CABG must be individualized based on both clinical and angiographic factors. When there is no

evidence of ongoing ischemia or hemodynamic instability, SCAD can be managed successfully with medical treatment alone (18). With conservative management partial or even complete angiographic resolution of coronary artery dissections has been observed after a follow up period of 2 months to 1 year. Coronary CT angiography can be used as an alternative imaging method for the assessment of angiographic resolution after medical treatment of SCAD. Medical management of SCAD is similar to the treatment of acute coronary syndromes. If a pronounced dissection persists in a major vessel after prolonged medical treatment, or in SCAD causing marked epicardial coronary flow impairment and/or ongoing ischemia, PCI or CABG should be considered. PCI with stenting can restore flow in the true lumen, relieving ischemia, and seal the dissection, preventing further expansion. Technical difficulties during PCI include advancing the guidewire in the true lumen rather than in the false lumen, and avoiding distal propagation of the intramural haematoma and dissection during stent delivery. The latter can be prevented by deploying the first stent with sufficient coverage of the distal border of the dissection. IVUS or OCT imaging can be used to confirm guidewire placement in the true lumen, evaluate the length of dissection and vessel size, assist in the correct positioning the first stent to deliver and assess stent apposition, and to seal the dissection at the end of the intervention. The clinical success rate of stenting in patients with SCAD is over 90%. Single vessel dissections of major coronary arteries are usually managed with PCI with stenting, while left main dissection, multivessel involvement, or failed PCI procedures are treated by CABG. In cases of spontaneous dissections involving a long coronary artery segment, CABG can be very challenging. The vessel wall may be fragile due to the underlying condition predisposing to dissection.

Prognosis

In-hospital mortality of SCAD is relatively low, with a mean rate of around 3 % (19). Patients who survive the acute phase have a good long term prognosis, with a very low recurrence rate of SCAD or acute coronary syndrome, and a 95% 2 year survival rate. Although outcome is in general good, the overall mortality in reported cases of the peripartum group is 38 % (20).

Conclusion

Incidence of SCAD is very low. SCAD occurs most often in young women (age <40 years). It occurs frequently in the peripartum period. Left anterior descending coronary artery is the most frequently involved vessel. Multivessel dissection is present in 20% of the cases. SCAD results from haematoma formation in the outer third of the vessel

wall and an intimal tear is only seldom observed. Early coronary angiography remains essential in the diagnosis of SCAD. Additional IVUS and OCT imaging provides very detailed information on the location and extent of the dissection. Patients with ongoing myocardial ischaemia must be treated with early revascularization (PCI or CABG). Patients without flow limiting dissection should be treated conservatively as spontaneous healing is possible. CT coronary angiography is useful in the follow-up of conservatively managed patients.

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