

CASE STUDY

Follow up after spontaneous coronary artery dissection: a report of five cases

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Abstract

Five cases of spontaneous coronary artery dissection (SCAD) are reported, three in women and two in men (mean age 44 years; range 28-65), all of whom suffered a myocardial infarction. Common risk factors for coronary artery disease were present in the two men; in the female group one patient was taking an oral contraceptive, one was in the postpartum period, and the third was a smoker. Only the three women received intravenous alteplase and their ejection fraction was normal; both men had impaired left ventricular function. Two patients had SCAD of the left anterior descending coronary artery and three of the right coronary artery. Only the two men had angiographic features of coronary atherosclerotic involvement. No patients required surgical revascularisation or percutaneous transluminal coronary angioplasty. At a mean follow up of 27 months (range 6 to 40) all patients were alive and all but one were asymptomatic.

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Keywords: myocardial infarction; coronary artery dissection; risk factors

Primary or spontaneous coronary artery dissection (SCAD) is a rare cause of ischaemic heart disease occurring predominantly in young, otherwise healthy women. It is often fatal and its incidence may be underestimated. To date the cases of a little more than 100 patients with this diagnosis have been reported. Most of these patients died as a direct result of the dissection. Only a few long

term survivors have been described.¹ The pattern and severity of presentation are variable and relate to the extent of dissection, its rate of development, and the vessel involved. Not unexpectedly dissection of the left main coronary artery or of the left anterior descending coronary artery (LAD) may have the most disastrous consequences including sudden death, extensive infarction, severe pump failure, and malignant arrhythmias. Pre-existing atherosclerotic disease may play a role in the clinical presentation, as may concurrent coronary spasm. Angiographic diagnosis of SCAD is extremely rare. This is a report of five cases of SCAD diagnosed by selective coronary arteriography from 2225 consecutive patients (0.2%) who underwent angiography at the department of cardiology of Rovigo, Italy, from January 1989 to August 1993.

Case report

Clinical data of patient population are summarised in the table.

CASE 1

A 28 year old woman was admitted to coronary care unit two hours after the onset of precordial pain. She had no history of previous cardiovascular disease, no collagen tissue disease, no trauma, and no risk factors for coronary artery disease. She had a full term uncomplicated spontaneous vaginal delivery six weeks before admission to our hospital. Physical examination was normal. The electrocardiogram showed ST elevation in leads V₂, V₃, and aVF. The echocardiogram showed an akinesis of the posterior segment of the left ventricular wall. The patient was treated with intravenous alteplase (100 mg). Serial electrocardiograms and serum enzyme

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Clinical characteristics of our cases with spontaneous coronary artery dissection

Case	Age	Sex	MI	TT	V	EF	Follow up
1	28	F	Inferior	+	RCA	0.72	A 6 months
2	42	F	Anterior	+	LAD	0.62	A 24 months
3	38	F	Anterior	+	LAD	0.71	A 38 months
4	65	M	Inferior	-	RCA	0.42	A 33 months
5	51	M	Inferior	-	LCA+RCA	0.26	24 months

MI, myocardial infarction; TT, thrombolytic therapy; V, vessel; EF, ejection fraction; F, female; M, male; A, asymptomatic; RCA, right coronary artery; LAD, left anterior descending coronary artery; LCA, left circumflex coronary artery.

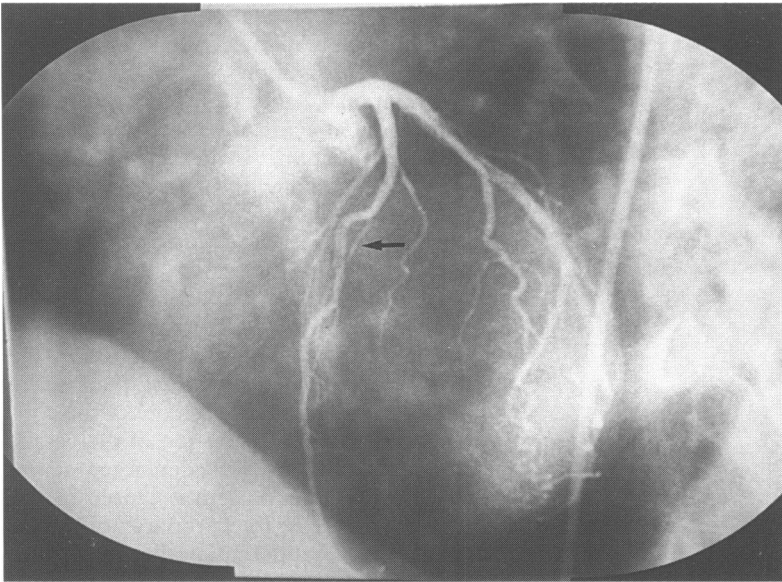


Figure 1 Left anterior oblique view with cranial angulation of the left coronary artery. Arrow points to intimal flap in the mid portion of the left anterior descending coronary artery.

levels showed acute myocardial infarction. During the following three days she had multiple episodes of chest pain with ST elevation in the inferior leads. She was given β blockers and intravenous glyceryl trinitrate and she improved symptomatically. Her later clinical course was uneventful. Thirteen days after myocardial infarction the patient underwent coronary and left ventricular angiography. Left ventricular angiography showed mild hypokinesis of the posterobasal and diaphragmatic left ventricular wall, with an ejection fraction of 0.72. Coronary angiography showed dissection of the right coronary artery. The dissection resulted in mild luminal narrowing of the

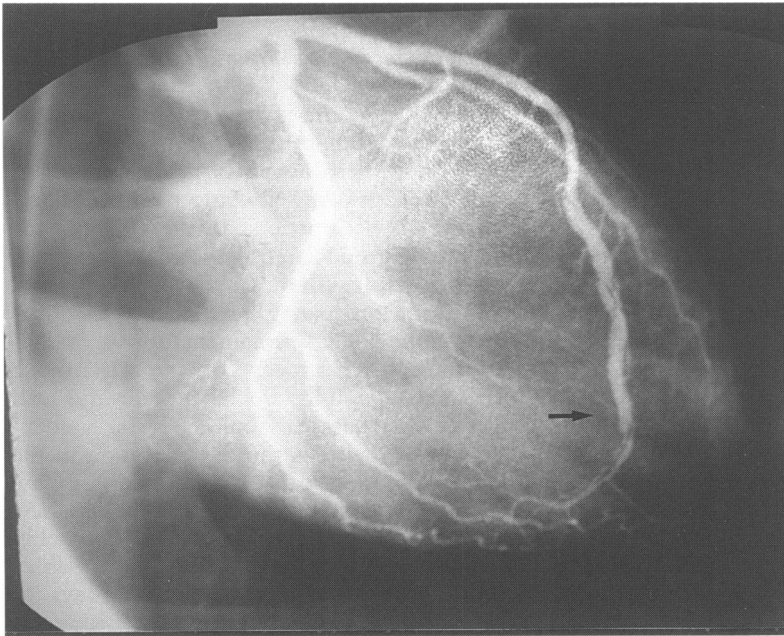


Figure 2 Left coronary angiogram (right anterior oblique) of patient number 3. Arrow points to the distal left anterior descending coronary artery where a dual lumen is present, separated by a flap-like spiral defect indicative of intimal dissection. The subintimal haematoma does not totally obstruct the lumen, as distal passage of dye occurs.

mid and distal segments of the artery. The patient no longer complained of chest pain, either at rest or during a submaximal exercise test. She was discharged on the 16th day and she was still asymptomatic at a six months clinical follow up.

CASE 2

A 42 year old woman who had previously been in good health experienced the acute onset of severe pain in the chest and left arm shortly after preparing breakfast at home. Three hours later she was admitted to our coronary care unit with an electrocardiographic and echocardiographic pattern of acute anteroseptal myocardial infarction. She had experienced three full term uncomplicated pregnancies, the last one three years previously; she was not taking oral contraceptives and had a normal menstrual status. She was a heavy smoker (20 cigarettes a day). She was given intravenous alteplase with improvement in her clinical state. However, serial electrocardiograms and serum enzyme levels showed evolution of acute myocardial infarction. Her clinical course was uneventful. Ten days after admission the patient underwent coronary angiography. Left ventriculography showed mild impairment of contractility in the anterior wall. The left coronary arteriogram showed a flap-like filling defect in the mid LAD (fig 1). The patient was discharged with medical treatment (β blockers and aspirin) after a negative submaximal exercise test. At a 24 month follow up she remained totally asymptomatic.

CASE 3

This 38 year old woman had experienced anginal pain while swimming. She was admitted to a hospital four hours later with evidence of acute anterior myocardial infarction and received intravenous alteplase. A month later she was referred to our cardiology department for further evaluation of her coronary artery disease. She had been taking an oral contraceptive at the time of her myocardial infarction, and had no further coronary risk factors. Physical examination was normal. A submaximal exercise test was negative. Left heart catheterisation revealed a normal left ventricular ejection fraction and apical akinesis. Coronary angiography showed a flap-like spiral filling defect indicative of an intimal dissection in the distal LAD, comprising the lumen (fig 2). Nevertheless anterograde flow was preserved. She was given medical treatment and was asymptomatic at a 38 month clinical follow up.

CASE 4

A 65 year old man was referred to our hospital for angiographic evaluation of his coronary artery disease. He had experienced a posterolateral myocardial infarction two years before. He had remained asymptomatic for the following period. A treadmill thallium test showed a posterolateral scar and an inferior reversible defect. The patient was a heavy smoker and had a history of hypertension and diabetes. Left ventriculography showed posterolateral

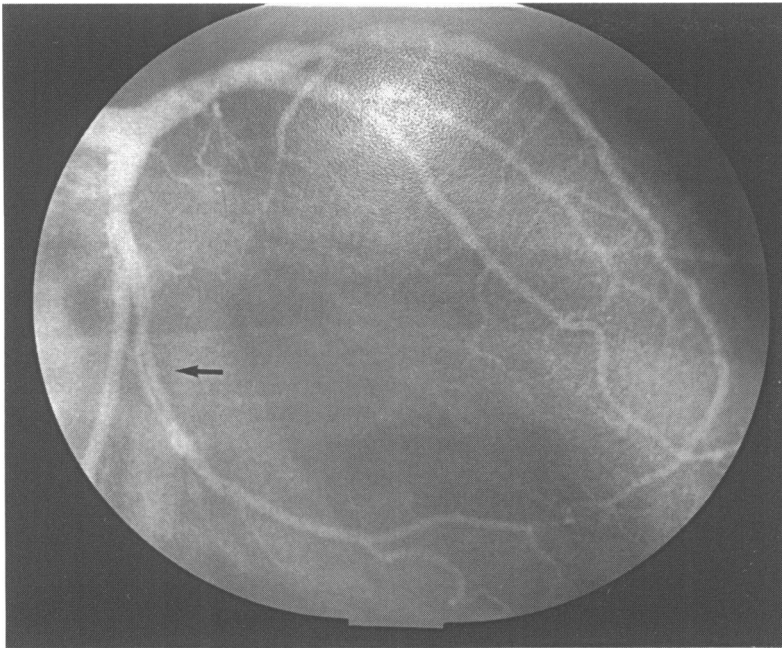


Figure 3 Right anterior oblique angiographic view of the left coronary artery showing (arrow) a serpiginous filling defect in the obtuse marginal branch of the left circumflex artery.

akinesis and mild anterolateral hypokinesis with an ejection fraction of 0.42. Coronary angiography revealed diffuse mild three vessel atherosclerotic disease with a long dissection of the right coronary artery in its mid and distal portions. We did not advise any surgical revascularisation and the patient was asymptomatic at a 33 month follow up.

CASE 5

This 51 year old man, who had several risk factors, had an inferior myocardial infarction two years before admission. In the years following this he experienced neither angina nor dyspnoea. After a positive exercise test he was referred to us for coronary angiographic evaluation. Left ventriculography and coronary angiography revealed severely impaired left ventricular function (EF 0.26) and dissections of the right coronary artery in its mid and distal portions and of the marginal obtuse branch of the left circumflex artery (fig 3). The antero-grade flow was preserved in both arteries. There were signs of diffuse atherosclerotic coronary artery disease with no evidence of severe focal stenosis. We decided to treat the patient medically. At a 24 month follow up he complained of effort dyspnoea (New York Heart Association grade II) and had ventricular arrhythmias which were treated with amiodarone.

Discussion

Spontaneous coronary artery dissection is extremely uncommon. Dissecting aneurysms of the coronary vessels are much more likely to be iatrogenic,² secondary to blunt chest trauma,³ or associated with an aortic dissecting aneurysm or with Marfan's syndrome. Definite physical features of the Marfan syndrome were absent in all our patients, and there was no history of previous trauma.

In most cases of SCAD the diagnosis is established on necropsy examination. A review of published reports shows that the usual clinical presentation of this condition was either sudden death or acute myocardial infarction followed in a short time by death. Though our patients had an acute myocardial infarction they survived the acute insult and all but one were asymptomatic at follow up.

The angiographic diagnosis during life of SCAD is very unusual.^{1,4} We defined dissection as the detection at coronary angiography of a radiolucent area within the lumen of the vessel, with or without contrast persistence within the dissection after wash out of the contrast from the remaining portion of the vessel. The lumen of the vessel was enlarged at the site of dissection; this pattern is caused by the presence of dye inside the coronary wall and is important in the differential diagnosis with intracoronary thrombus.

SCAD typically occurs in young, healthy women. In a series reviewed in 1990 by Cocco *et al*,⁵ 74 out of 97 cases were women, with an average of 39 years. It is striking that 24 of 74 patients were in the puerperium and one had been taking an oral contraceptive for the last three years. Approximately 50% of patients with a coronary artery dissection die suddenly and many others (18–20%) will die within a few hours. The clinical presentation is otherwise similar to that of patients with atherosclerotic coronary artery disease, although the average age is younger and risk factors may be absent. SCAD may be recurrent;⁶ it may also heal spontaneously.⁷ Van Der Bel-Kahn⁶ reported the case of a 40 year old man in whom a dissection of the obtuse marginal artery had apparently healed, but who died at a later date of a second dissection of the LAD.

Himbert *et al*⁷ described progressive angiographic healing of a spontaneous dissection of the left main coronary artery. The LAD is the most commonly involved artery (52.5%),⁵ followed by the right coronary artery (24%), the left main stem (13.5%), and the left coronary artery (2%).

Various hypothesis have been offered to explain the aetiology of SCAD. Hypertension does not seem to be a risk factor, since it is usually absent.⁵ Any unifying theory of coronary artery dissection must explain the remarkable predilection for women, especially during the puerperium. Changes in the arterial wall during pregnancy have been well documented and include fragmentation of reticulum fibres, hypertrophy of smooth muscle cells, and alterations in the protein and acid mucopolysaccharide content of the media.⁸ The current belief is that degeneration of the ground substance causes a weakening of the tunica media of the vessels during pregnancy; the effect of straining during labour and delivery may initiate intimal rupture, with subsequent haemorrhage into the media days or weeks later. SCAD has occurred in men, however, thus indicating multifactorial aetiology. Angiitis has been implicated in the pathogenesis of SCAD. Robinowitz *et al*⁹ found in a necropsy study that their eight patients and 31

(43%) of the previously reported necropsy cases had an eosinophilic infiltrate in the adventitia of the involved coronary artery. They postulated that proteins isolated from the granules of eosinophils may have damaged the collagen, elastin, or smooth muscle cells of the coronary artery, resulting in dissection.

Two of our patients showed both SCAD and atherosclerotic disease. Berger *et al*¹⁰ showed that in patients with areas of atherosclerotic injury there is increased density of vasa vasorum and marked neovascularisation. Newly formed vessels are known to be relatively fragile and might form the initial focus of haemorrhage.

The prognosis of the patients who survived to the acute insult has not been defined. De Maio *et al*¹ followed up 27 patients for 1.5 to 144 months (mean 38). Twenty two survived the acute event (82%) and have done well, all but one remaining asymptomatic. All our five patients are alive and four are asymptomatic at a mean follow up of 27 months (range 6–40). At a 24 month follow up patient number 5 complained of effort dyspnoea and had ventricular arrhythmias.

It is not clear whether SCAD should be treated medically or surgically. Medical treatment may have a role. Vacek and McKiernan¹¹ reported that a patient with a catheter induced left coronary dissection was successfully managed initially with intracoronary streptokinase. Ramamurti *et al*¹² gave streptokinase to a patient before the diagnosis of dissection was suspected. Three of our patients received alteplase and their ejection fraction was preserved. This treatment may be effective in lysing thrombi in the false lumen, thereby allowing the true lumen to re-expand. Glyceryl trinitrate may be useful in releasing spasm sometimes associated with dissection.¹³

None of our patients required surgical or PTCA treatment. Antoniucci and Diligenti reported a patient with SCAD of three vessels who received coronary bypass grafting.¹⁴ Thayer *et al*⁴ recommended bypass grafting for all patients with SCAD. Vicari *et al*¹⁵ described surgical repair of SCAD by extrusion of an intraluminal haematoma. Gonzales *et al*¹⁶ described a case treated successfully with PTCA. The authors pointed out that the risk of coronary perforation would seem to be significant, as the guidewire and angioplasty balloon were passed blindly into a vessel that

was already damaged and presumably weakened. We agree that the indications for bypass grafting or coronary angioplasty are the same as for any other case of severe narrowing of an epicardial coronary artery with clinical evidence of myocardial ischaemia.

Our series shows that patients with SCAD who have developed an infarct in the absence of fixed and severe stenosis do well with medical treatment. The indications for surgery are the same as those for patients with fixed atherosclerotic disease: left main disease, three vessel disease, or symptoms refractory to medical treatment.

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