MULTIVESSEL SPONTANEOUS CORONARY ARTERY DISSECTION IN A YOUNG WOMAN

Sandra Alban, Elvis Brscic, Salvatore De Salvo, Alessandro Decio, Paolo Russo, Alessandro Alberti*

Invasive and Acute Cardiology Unit Villa Maria Pia Hospital, Turin, Italy
*Centro studi IVUS, Clinica Cellini, Turin, Italy

Case report

The patient was a 38-year-old woman with negative family history for ischemic cardiopathy, not hypertensive, diabetic, smoker or dyslipidemic. She had no medical history for drug addiction or estroprogestinic therapy. She had a physiological pregnancy when 21 years old and regular menstrual periods. Her last menstrual period was 16 days before the event. Her surgical history consisted of thoracic trauma when 6 years old and bilateral saphenectomy. She suffered from periodic cluster headache, but no connective tissue pathology.

The patient was referred for anterior-septum STEMI, treated by systemic trombolysis (accelerated rTPA) one hour from the onset, with clinical and electrocardiographical reperfusion criteria. The patient also received IV heparin, β-blockers, nitrates and IV morphine, ASA and ACE-inhibitors. Echocardiography demonstrated LVEF of 40%, anterior interventricular septum akinesia, anterior and lateral wall hypokinesia, mild mitral insufficiency, normal pulmonary artery systolic pressure (20 mmHg), a regular aorta, and no pericardial effusion. Six hours after thrombolysis, angina pectoris recurred with repeated episodes, transient modifications of ECG, increased ischemic area at echocardiography (EF 30%) and progressive haemodynamic instability.

Urgent coronary angiography was carried out through the right femoral approach and was completed by intracoronary ultrasound examination (IVUS) study; it showed spontaneous coronary dissection of the proximal and medium parts of the LAD (Fig 1).

In the same setting, we successfully implant a direct stent (bare metal stent) in the LAD and an intra-aortic balloon counterpulsation (IABP). The patient rapidly improved, symptoms resolved with satisfactory haemodynamic balance; IABP was removed after 24 hours without complications. Peak creatin-kinase (CK)/CK-MB value was 3285/296 UI/l at 7 hours from hospital admission and before the angiography, with following gradual and constant reduction of myocardial cytolysis index. ECG evolved in anterior-septal and lateral necrosis with antero-lateral ischemia. At control echocardiography carried out after mechanical support removal, left ventricular ejection fraction was 50%, with apical akinesia and hypokinesia of periapical segments of anterior wall and septum, without pericardial effusion.

The subsequent course was uneventful; the patient was discharged on the 5th day, asymptomatic and in good haemodynamic balance, on ASA (160 mg/die)-Ticlopidin (500 mg/die) for 15 days, β-blockers and ACE-inhibitors.
Few weeks after returning home, the patient was pregnant and decided to abort, in the manner provided for by the law. She underwent cardiac work-up which demonstrated normalization of contractile function of left ventricle at echocardiography and negative stress test for symptoms and ischemia. After 6 months, angiography was negative for coronary stenosis (Fig. 2).

After 2 years from the infarction, there was an episode of chest pain at rest, without ECG modifications, enzymatic movement and/or massive alterations of segment kinesis at echocardiography. Angiography confirmed good outcome of previous revascularization of LAD but showed a critical lesion of medium tract of right coronary, followed by distal sub occlusion. IVUS study of the vessel demonstrated spontaneous dissection of the tract appeared stenotic at angiography, and an image of a distal haematoma, partially occluding the vessel (Fig. 3).

Consequently, we performed percutaneous revascularization of the right coronary by direct stenting, obtaining normalization of anterograde flow.

Gynaecological examination showed uterine fibromyomatoses (two intramural sub serous myomas 34x33 mm) and the presence at haemocoagulative and autoimmune screening of low level anti-nucleus antibodies (1:80 at the granular fluorescence) and anti-Scl-70 (not confirmed at the next control). The patient refused further immunological tests.

Six months after the second percutaneous coronary revascularization, the patient underwent a control coronary angiography because of a new episode of chest pain. Good outcome of the previous procedure was confirmed. However, important vessel catheter spasm of the right coronary obliged verapamil therapy to obtain control of symptoms; it wasn’t possible to use nitrates because of worsening headache.

After 5 years, the patient is asymptomatic in medical therapy (ASA 160 mg/die, verapamil 240 mg/die, atorvastatin 20 mg/die), she has normal professional and recreational activity.

Discussion

Even though atherosclerosis is the most common cause of coronary disease, other conditions can be responsible for the pathology; among them, coronary spontaneous dissection is one of the most deceitful. Evaluated by Nishikawa as low as 0.28%, its incidence is underestimated since it is frequently detected only at post-mortem examination.

We defined coronary dissection as the separation of the...
media from the other vessel layers by a haematoma, pushing the internal-medium wall toward the vessel lumen, causing a more or less severe limitation of the blood flow.

The coronary dissection can be spontaneous or secondary to an aortic dissection, Marfan's Syndrome, thoracic traumas, physical efforts\(^7\), or to iatrogenic causes (cardiovascular surgery, coronarography or coronary angioplasty). About 220 cases have been described in literature\(^8\); most involved are women (70-80%), 40 year-old on average; in 1/3 of the cases the event is related with last pregnancy trimester, first post-partum period or estroprogestinic therapy\(^9,10\).

The most frequently involved artery is the LAD in women, the right coronary in men.

A recent classification of spontaneous coronary dissection\(^11\) divides them in 3 groups. The first is made of young women in parturient period and it represents the most common group in literature. The second one is made of patients affected by coronary atherosclerosis; in these patients dissection is just a phenomenon associated with the development of the plaque. The third group, defined as idiopathic, is made of patients in whom coronary dissection is the only evidence of lesion.

In pregnancy associated forms, it is possible that the hormonal modifications alter the synthesis of the acid mucopolysaccarids and the proteins which build up the media of the coronary vessel, as we can see in aorta, weakening the vessel wall and promoting the intimal rupture\(^12,13\). In the same way, hyperestrogenic state was considered responsible for coronary fragility in female patients who assumed estroprogestinic therapy. Furthermore, coronary spontaneous dissection was found in patients with altered collagen synthesis, as in Ehlers-Danlos syndrome\(^14\).

The patient we encountered has got some peculiar characteristics. The absence of common risk factors for coronary pathology (familiar history for ischemic cardiopathy, hypertension, dyslipidemia, diabetes mellitus, and current smoking) suggests an idiopathic form of coronary dissection. On the other hand, pregnancy (even if at an early stage) at the time of the first coronary event and uterine fibromatosis correlated to a hyperestrogenic state at the time of the second coronary event suggest a possible role for sexual female hormones. Besides, the positive research for anti-nucleus and anti-Scl70 antibodies, even if at a low level, can also suggest an autoimmune pathology of connective tissue with vascular involvement.

In the literature several cases are described in which dissection involves many important coronary branches simultaneously; but it is quite rare that the dissection involves two different vessels in different period of time, as we observed in our patient.

Finally, intracoronary ultrasound examination is fundamental to diagnose unusual lesions such as the spontaneous coronary dissection and to provide therapeutic indications to treatment.

Spontaneous coronary dissection should be included in differential diagnosis of acute coronary syndrome, mainly in young women; only early coronary angiography can confirm the suspected diagnosis and allow the appropriate treatment.

The optimal management of spontaneous coronary artery dissection is uncertain for the limited clinical experience and the lack of controlled trials. At this time, given the unclear pathophysiology, there are no guidelines on optimal therapy.

Reported treatment options include medical therapy (heparin, aspirin, clopidogrel, glycoprotein IIb/IIIa inhibitors, statin, β-blocker, nitrates and ACE inhibitor), thrombolysis (but in some cases have been shown to potentially worsen the dissection), percutaneous transluminal intervention with stenting and coronary artery bypass surgery\(^15,16\). Treatment should be highly individualized on a case-by-case basis. When the patient is hemodynamically stable, without sign of ongoing ischemia, an initial treatment strategy of medical management can delay the need of mechanical intervention. Patient with active ischemia or failed medical management require early percutaneous or surgical revascularization.

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Bibliography


Address for correspondence:
Sandra Alban
Villa Maria Pia Hospital - Str. Mongreno, 180 - 10132 Torino
Tel. +39.011.8967334 - E-mail: sandra.alban@email.it