Spontaneous left main coronary artery dissection

A rare cause of acute coronary syndrome

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ABSTRACT

يعد تسلخ الشرايين التاجية التلقائي سبب نادر لمتلازمات الشريان التاجي الحادة والموت المفاجئ. تسلخ الشريان التاجي الرئيسي أمر نادر الحدوث ويمثل ما نسبته 9% من جميع الحالات. تصيب هذه الحالة الإناث في الغالب في سن ما قبل انقطاع الطمث، و تحدث ثلث تلك الحالات أثناء فترة الحمل و الفترة المحيطة بالولادة. ربما يتم إغفال التشخيص السريري أحياناً حيث أن المرضى غالباً ما يكونون في سن الشباب و ليس لديهم عوامل الخطر بالنسبة لأمراض الشرايين التاجية. نستعرض في هذا التقرير حالة مريضة تبلغ من العمر 42 عام أصيبت بتسلخ الشريان التاجي الرئيسي بعد الولادة، مع مناقشة وجيزة لهذه الحالة المرضية و علاجها.

Spontaneous coronary artery dissection is a rare cause of acute coronary syndrome and sudden cardiac death. Involvement of the left main coronary artery is uncommon accounting for 9% of all cases. The condition commonly affects premenopausal females with about one third of the cases occurring during pregnancy and the peripartum period. The diagnosis may occasionally be overlooked as the patients are often young and have no risk factors for coronary artery disease. The clinical presentation and interesting angiographic findings of a 42-year-old female patient with left main coronary artery dissection are described along with a brief discussion of the pathogenesis and management of the condition.

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Spontaneous coronary artery dissection (SCAD) is an uncommon but important cause of acute myocardial infarction in the young. It commonly affects healthy females during pregnancy and the postpartum period. A high index of suspicion is required for prompt diagnosis and proper management as clinical instability and sudden death are potential complications. The following is a description of a patient who presented with this condition, along with a discussion of the pathogenesis of the lesion and therapeutic considerations.

Case Report. A 42-year-old previously healthy woman presented to her community hospital with sudden retrosternal chest tightness and severe shortness of breath. The patient had a normal delivery 2 days before and delivered a full term healthy boy. Pregnancy was uneventful. Her medical history was unremarkable and she did not have risk factors for coronary artery disease (CAD). On examination, she was dyspneic and orthopneic. The respiratory rate was 23/minute; the blood pressure was 120/80 mm Hg and the pulse rate was 80/ minute. The heart sounds were soft and no murmurs were detectable. Chest examination revealed bilateral widespread fine crackles. Chest x-ray showed increased cardio-pericardial silhouette and interstitial pulmonary edema. Twelve-lead electrocardiogram (ECG) showed minimal ST segment elevation in the precordial leads and first set of cardiac enzymes was normal. The patient was initially thought to have peripartum cardiomyopathy and was managed with nitrates and diuretics. Repeat ECG few hours later showed marked ST segment elevation in the precordial leads, and leads I and aVL. Extensive anterior ST-elevation myocardial infarction (STEMI) was diagnosed and she was treated with aspirin and thrombolytics, but there were no signs of reperfusion. She remained clinically stable and was transferred to our tertiary care center few days later for coronary angiography. Upon arrival to our center, she was free of pain. The blood pressure was 99/70 and the pulse rate was 120/ minute.

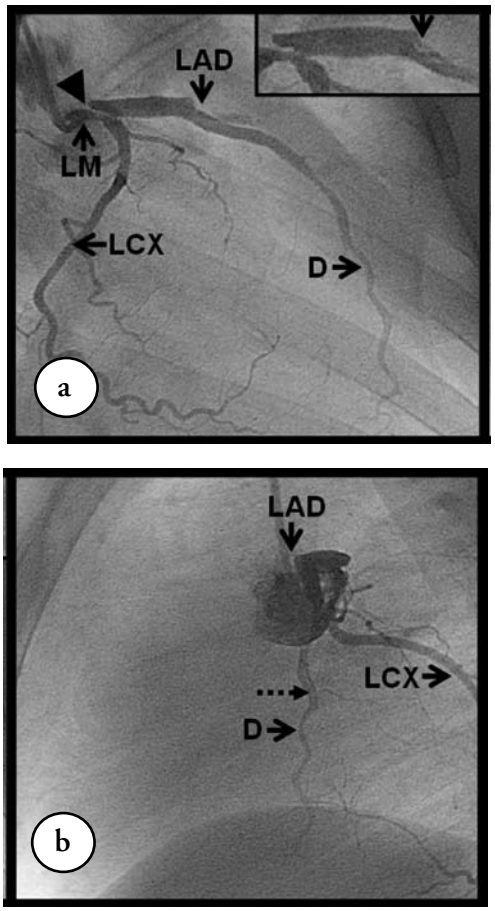

Figure 1 - Angiogram of the left coronary system. **a**) Right anterior oblique and **b**) left lateral (b) projections. Dissection of the left main (LM) coronary artery (black arrow) with almost total occlusion of the left anterior descending artery (LAD) is seen. The dissection also extends to a large diagonal (D) branch (dotted arrow). The dissection is compromising the ostium of the circumflex artery (LCX).

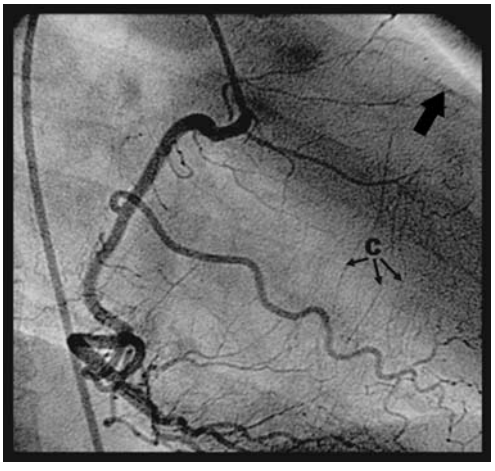

Figure 2 - Right anterior oblique projection of the right coronary artery (RCA), which appears angiographically normal. The left anterior descending artery (large arrow) is partially filling retrogradely by collaterals (C) from the RCA.

The Jugular venous pressure was 5 cm above the sternal angle and she had crackles over both lung bases. A 12 lead ECG showed a completed anterior infarction with Q waves and loss of the R wave in the precordial leads. Troponin I was minimally elevated (0.47 µg/L, normal <0.04 µg/L) and creatine phosphokinase (CPK) was normal. Coagulation profile, kidney function, and liver enzymes were normal. Total cholesterol was 5.8 mmol/L (normal 3.2-5.2 mmol/L). Antinuclear antibody, and antiphopholipid antibodies were negative. Transthoracic echocardiography revealed a dilated left ventricle with an akinetic anterior wall and apex. The ejection fraction was 25% and there was mild mitral regurgitation.

Coronary angiography revealed spontaneous dissection and luminal narrowing of the left main coronary artery (LMCA). The left anterior descending artery (LAD) was almost totally occluded. The dissection extended to a large first diagonal and compromised the ostium of the left circumflex (LCX) artery (**Figures 1a & 1b**). The right coronary artery (RCA) was normal and was attributing collaterals to the LAD (**Figure 2**). Left ventriculogram revealed anterior wall akinesis and

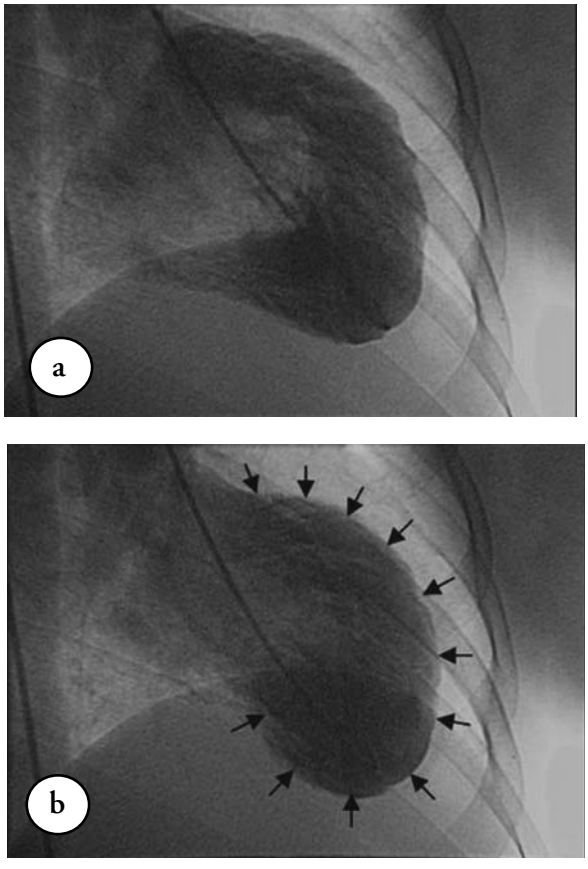

Figure 3 - Left ventriculogram in right anterior oblique position, **a**) diastolic and **b**) systolic views showing akinesis of the anterior wall and dyskinesis of the apex (arrows).

dyskinesis of the apex (**Figures 3a & 3b**). The patient was referred for emergency bypass surgery.

Discussion. Spontaneous coronary artery dissection is a rare cause of acute coronary syndrome (ACS) and sudden cardiac death (SD). It is observed in 0.1 % of patients undergoing cardiac catheterization. This figure however may underestimate the true incidence as a large number of SCAD lead to SD without being diagnosed.[1] Females outnumber males constituting 63% of the cases. One third of affected females were either pregnant or in the postpartum period. Mean age at presentation is 46 years (range 21-82). Involvement of the LMCA and multivessel dissection is uncommon, accounting for less than 10% of the cases.[2] Presentation includes a broad clinical spectrum ranging from stable angina pectoris to cardiogenic shock due to extensive myocardial infarction or even sudden cardiac death. Syncope and presentation with congestive heart failure have also been reported.[1,2] The etiology of the condition remains largely unknown. Reported associations include physical exertion, hypertension, cocaine abuse, oral contraceptive use, fibromuscular dysplasia and Ehlers-Danlos syndrome.[2] The higher incidence of SCAD during pregnancy and the peripartum period is a well recognized phenomenon. Two mechanisms have been implicated to cause SCAD during pregnancy and the peripartum period: mechanical vessel failure secondary to altered endocrine status during pregnancy; and increased hemodynamic stress during delivery leading to rupture of the thin walled vasa vasorum especially in the presence of medial alterations.[3] Among the endocrine factors, estrogen and progesterone have been linked to disruption of the media and medial necrosis through increasing the release of matrix metalloproteinases (MMPs), reducing the normal corrugation of elastic fibres, decreasing the acid mucopolysaccharide ground substance and promoting smooth muscle cell hyperplasia. Relaxin is another pregnancy hormone that also increases several MMPs.[4] It also has a potent chronotropic and inotropic effects which may increase the risk of dissection.[3] These pregnancy associated hormonal changes have also been associated with increased risk of aortic dissection.[4] The underlying pathology in SCAD is an intramural hemorrhage leading to compression and occlusion of the vessel lumen. The dissection plane is in the outer media or between the media and adventitia. Rupture of vasa vasorum as a result of the degenerated media appears to be the source of the hematoma in SCAD.[1] An intimal flap is rarely observed. Other encountered abnormalities include eosinophilic periadventitial inflammation.[1,2] There are 4 salient features of SCAD: female patient, young age, presentation with an acute myocardial infarction, and presentation in the peripartum period.[5] A high index of suspicion is required for early diagnosis. Patients with suspected spontaneous coronary artery dissection should be referred for urgent coronary angiography. Thrombolytics are better avoided as their use has been associated with increase in the hematoma size, propagation of the dissection and clinical deterioration.[1] The diagnosis is usually made at the time of cardiac catheterization. Angiographic findings of dissection include coronary occlusion, thrombus formation, presence of an intimal flap, persistence of contrast dye in the false lumen and variability in the size of the coronary lumen during the cardiac cycle. The last finding results from pressure differences in the true and false lumens during the phases of the cardiac cycle.[6] Occasionally, recognition of the dissection may be difficult on cardiac catheterization. Intravascular ultrasound (IVUS) can confirm the diagnosis in selected cases; however, it is important to be aware of the risk of wiring the false lumen.[1] Multidetector computed tomography is another useful diagnostic modality and is of benefit for follow up of cases.[7] Spontaneous dissection of the LMCA requires emergency revascularization as the area of the myocardium involved or at risk is usually large. Clinical instability is also a major concern. Patients who have completed the acute event and have no evidence of ongoing ischemia and no significant stenosis on coronary angiograms may do well with medical therapy temporarily; however, the majority eventually develop recurrent angina, subsequent infarction or sudden death due to dissection extension.[8] Left main coronary artery dissections represent complex bifurcational lesions as involvement of the LAD, LCX or both is common.[8] Although coronary stenting may be a reasonable option for patients with single vessel dissection, it is better avoided in the case of LMCA and mulivessel dissection.[1,2] Complications such as extension of the hematoma, and the need for multiple stents to achieve revascularization have been encountered with percutanous interventions. Resolution of the intramural hematoma with time, has been reported to cause stent non-apposition.[9] The objectives of surgery are to restore flow beyond the obstruction and to minimize the chance of further dissection. Anastomosis of grafts should be performed in sections without or with minor atherosclerosis. It may not be feasible to go distal to the dissection as the vessel may be too small, which may reduce the likelihood of long-term graft patency. If the dissection involves the anastomotic site, the layers of the vessel should be re-approximated during the anastomosis. When the anastomosis is performed in a non-dissecting portion, a false lumen remained in most cases, which did not affect the patency of the bypass.[10] Occasionally, it may be technically challenging to ensure bypass in the true arterial lumen. Use of both antegrade

and retrograde blood cardioplegia when opening the artery may facilitate identifying the true lumen.[8] The prognosis of patients with SCAD overall has improved due to early diagnosis and aggressive management in the current era. The outlook for those who survive the initial event is good with mortality of 18% over a mean follow-up of 41 months.[2] It is estimated however, that in up to 50% of patients, a second dissection develops within 2 months of the initial event.[1] In our patient's case, in view of the possibility of dissection recurrence, and considering her severe LV dysfunction, future pregnancy was considered to carry a significant risk and was discouraged.

In conclusion, SCAD is a rare but important cause of ACS and SD that commonly affects otherwise healthy premenopausal females. Once the condition is suspected, coronary angiography should be performed immediately. This will be of importance in both establishing the diagnosis and guiding further management.

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