

Spontaneous coronary artery dissection and aortic dilatation presenting concomitantly: a case report

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Introduction

Spontaneous coronary artery dissection (SCAD) is defined as a non-traumatic, non-iatrogenic, non-atherosclerotic separation of the coronary arterial walls, creating a false lumen. The space created is filled with an intramural haematoma (IMH) that compresses the true arterial lumen, decreasing anterograde blood flow. Spontaneous coronary artery dissection is commonly associated with small and medium sized extracoronary vascular abnormalities.

Case presentation

This case report describes a case of SCAD presenting as an acute coronary syndrome together with aortic dilatation requiring aortic valve and aortic root replacement.

Discussion

Despite the fact that SCAD and aortic dilatation share common aetiologies, this is the first case to our knowledge describing severe aortic dilatation and SCAD presenting concomitantly. This case highlights the importance of confirming the diagnosis of SCAD with intravascular imaging and of investigating for extracoronary arteriopathies.

Keywords

Spontaneous coronary artery dissection • Aortic dilatation • Cystic medial necrosis • Cystic medial degeneration • Aortic regurgitation • Case report

Learning points

- Spontaneous coronary artery dissection can present as an acute coronary syndrome, and the diagnosis can be confirmed using intravascular ultrasound or optical coherence tomography.
- Spontaneous coronary artery dissection frequently presents in conjunction with other forms of arteriopathy which should be sought actively using dedicated protocols.
- Spontaneous coronary artery dissection can frequently be managed conservatively but the associated arteriopathies may need treatment.

Introduction

Spontaneous coronary artery dissection (SCAD) consists of the creation of a false lumen between the layers of the coronary wall. This separation can occur between the intima and media or between the media and the adventitia and is filled with an intramural haematoma (IMH) that compresses the true arterial lumen, decreasing anterograde blood flow.¹ Spontaneous coronary artery dissection is associated with small and medium sized extracoronary vascular abnormalities in 69% of cases.² Despite the fact that some causes of SCAD are also causes of aortic dilatation, a case involving aortic dilatation and SCAD concomitantly has not previously been described. Conditions that may lead to joint SCAD and aortic dilatation may

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include fibromuscular dysplasia (FMD), connective tissue disorders (CTD), and cystic medial necrosis (CMN).

Timeline

Day Events

- | Day | Events |
|-----|--|
| 1 | Presentation with acute onset chest pain, electrocardiogram changes and a troponin rise and a computed tomography aortogram showing aortic dilatation without evidence of dissection |
| 2 | Coronary angiogram showed a stenosis in the 2nd obtuse marginal which was shown by optical coherence tomography to be a spontaneous coronary artery dissection with an intramural haematoma. |
| 60 | Repeat coronary angiogram showed resolution of the coronary dissection. |
| 120 | Aortic root and aortic valve replacement |

Case report

Patient information

A 67-year-old female patient was admitted with a 3-h history of acute onset chest pain radiating to the interscapular region. On examination, she was haemodynamically stable with normal heart sounds and a normal respiratory examination. She was hypercholesterolaemic, obese and had a family history of aortic disease (her brother had been diagnosed with dilatation of the ascending aorta and subsequently died suddenly; her niece had aortic coarctation repair and has a bicuspid aortic valve).

Diagnostic assessment

At presentation minimal ST depressions in V3–V4 were evident on the electrocardiogram (ECG), troponin I was significantly elevated

[99.0 ng/L (<0.01 ng/mL)] as was creatinine kinase [1769 U/L (22–198 U/L)]. A computed tomography (CT) aortogram performed due to a wide mediastinum on chest X-ray showed an elongated and dilated aorta (5.2 cm × 5.1 cm) without evidence of dissection. An echocardiogram showed a moderately dilated left ventricle (LV) with reduced global function (ejection fraction 35%), akinesia in the basal and mid lateral segments, inferior hypokinesia, and moderate aortic regurgitation (Supplementary material online, Video S1). An aortogram confirmed CT findings of aortic dilatation (Figure 1A and Supplementary material online, Video S2). A coronary angiogram revealed a 20 mm long severe stenosis (70%) in the 1st obtuse marginal which featured abrupt changes in calibre from the surrounding normal artery and unimpaired distal flow (Figure 1B, Supplementary Video 3). Since there were no other coronary lesions in other parts of the coronary tree and in view of the generalized tortuosity of the coronaries, SCAD was suspected. The suspicion of type 2 SCAD was confirmed with optical coherence tomography (OCT) which revealed an 18 mm long IMH causing a true minimal luminal area of 1.6 mm² with an intimal tear in its distal aspect (Figure 2A, Supplementary material online, Video S4).

Interventions

Conservative management of the SCAD with β -blockers (nebivolol 5 mg daily) was opted for and an aortic root replacement was planned electively, but after 2 months she was readmitted with chest pain similar in nature to the index presentation. A repeat CT thorax showed no interval change in the aorta and a repeat angiogram showed normalization of the 1st obtuse marginal's calibre (Figure 1C, Supplementary material online, Video S5). A repeat OCT confirmed resorption of the IMH and normalization of luminal calibre (Figure 2B, Supplementary material online, Video S6). Two months later she underwent an aortic valve replacement with a biological valve and substitution of the ascending aorta with a 30 mm hemashield prosthesis. Histological examination of the ascending aorta showed myxoid degeneration of the media.

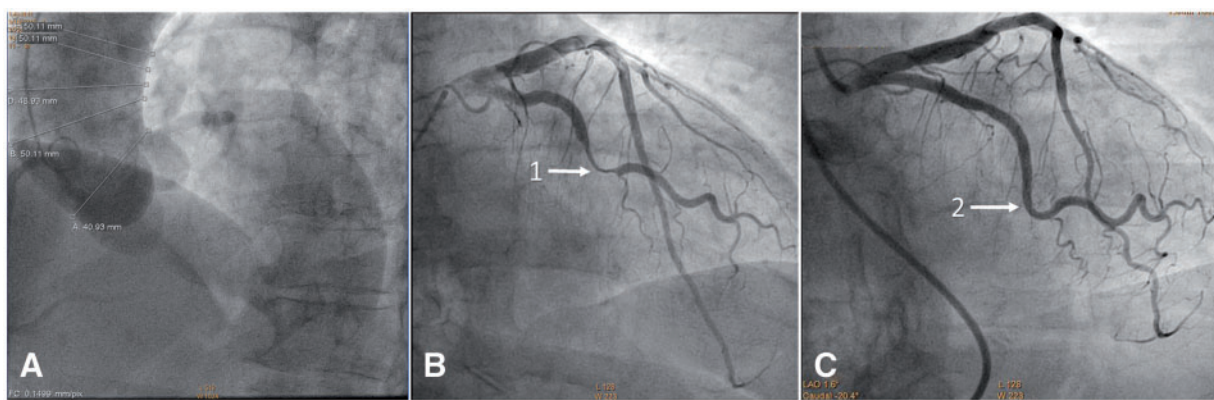


Figure 1 Aortogram showing aortic dilatation involving the whole aorta and tortuosity of the descending aorta (A). Type 2 Spontaneous coronary artery dissection (1) involving the first obtuse marginal on presentation angiogram (B). Complete angiographic resolution (2) can be observed in the repeat coronary angiogram after 2 months (C).

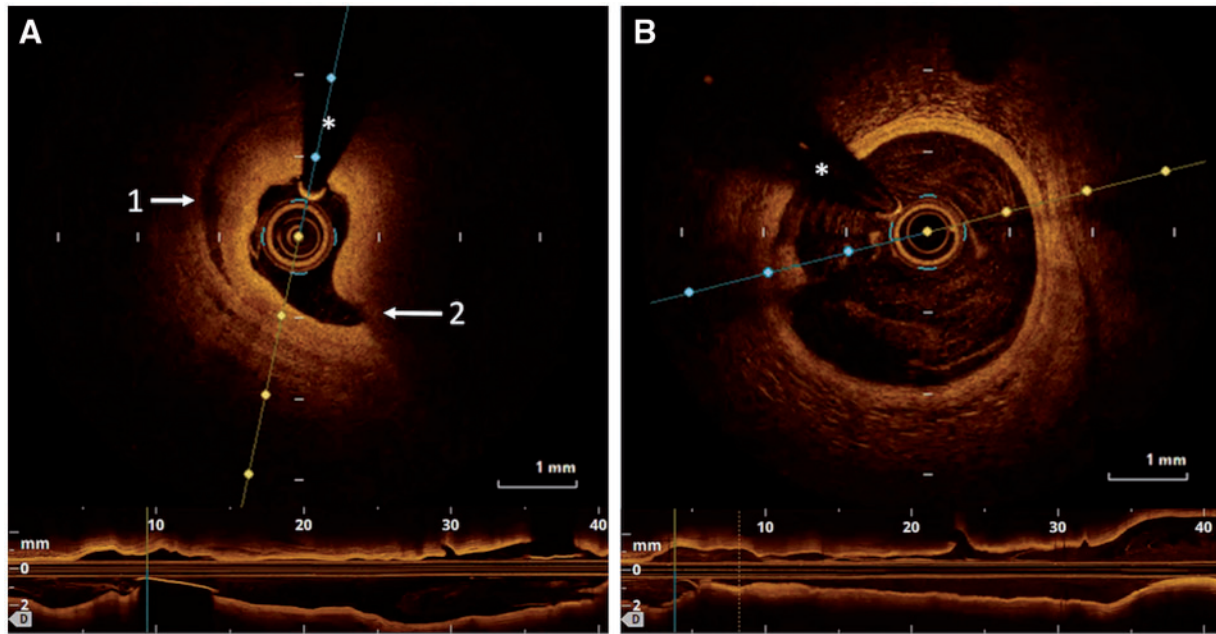


Figure 2 An intramural haematoma (1) and possible dissection flap (2) encroaching on the lumen can be observed in initial optical coherence tomography performed at presentation (A). The second optical coherence tomography (B) performed after 2 months shows complete resolution of these changes. *Guidewire.

Follow-up and outcomes

Based on the histological examination a diagnosis of CMN was made. At 3-month follow-up, she has remained asymptomatic in New York Heart Association (NYHA) Class 1 with no other cardiovascular events.

Discussion

Spontaneous coronary artery dissection frequently occurs in patients with a predisposing arteriopathy such as FMD, pregnancy, CTD, or systemic inflammatory conditions. In patients with a predisposing arteriopathy, a precipitating stressor usually leads to the development of SCAD. Such stressors include intense exertion, the valsalva manoeuvre, pregnancy, or recreational drug abuse.¹ Whether SCAD is an inheritable condition is still debated. Heritability was described recently in a few families which were found using social networking;³ however, single-centre registries have not found features strongly suggestive of a heritable disorder.⁴

Large artery involvement in the form of thoracic and abdominal aortic tortuosity and undulation have been described in 10% of SCAD cases investigated using a complete CT protocol.² However, dilation of the aorta as seen in this case has not been previously described. It is reasonable to postulate that the aortic dilation and coronary dissection were secondary to a common condition weakening both small and large sized arteries.

Fibromuscular dysplasia is the most frequently observed arteriopathy and is found in 72–86% of SCAD cases.¹ Fibromuscular dysplasia usually involves small and medium sized arteries¹ but has been reported to involve the aorta in over 30 cases, manifesting as aortic coarctation, abdominal aneurysms, or aortic hypoplasia with only two reports of aortic dissection.⁵

Other conditions in which aortic dilation and SCAD have been described albeit not in the same patient include Marfan's and Ehlers–Danlos Type IV and CMN/degeneration.⁶ Cystic medial necrosis is usually considered a disorder of large arteries, particularly the aorta. Cystic medial necrosis consists of loss of smooth muscle cells and fragmentation of elastic fibres with the appearance of cystic spaces filled with mucopolysaccharides. Cystic medial necrosis is a common pathological finding in various conditions rather than a distinct condition.⁷ In fact, CMN is a common histological finding in CTD including Marfan's, Ehlers–Danlos syndrome Type IV, familial thoracic aortic aneurysm, but it also occurs normally with aging and is accelerated by hypertension.⁸ Histology cannot distinguish between CMN changes brought about by these CTD and those found normally with aging.⁸ Interestingly many cases of SCAD that have been microscopically examined have also shown features of CMN in both the dissected and non-dissected segments.⁹ Besides coronary dissections CMN has also been associated with aneurysms of the coronary arteries and spontaneous dissection of the internal thoracic artery.⁷ Other causes of aortic dilatation such as syphilitic aortitis, giant cell aortitis, non-specific aortitis, senile degeneration of the aorta, trauma, yaws, inflammatory diseases such as bacterial or fungal aortitis, takayasu arteritis, giant cell arteritis, and idiopathic dilatation of the aortic root have not been described in association with SCAD.^{8,10} Since the histological examination of the ascending aorta showed myxoid degeneration it is plausible that in our case SCAD and aortic dilatation were both secondary to CMN.

This case demonstrates the importance of suspecting the diagnosis of SCAD in patients with tortuous arteries and isolated stenosis. This suspicion should be confirmed with intravascular imaging and in confirmed cases extracardiac arteriopathies should be looked for.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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