CASE REPORT

ST segment elevation myocardial infarction of a rare aetiology: an unexpected diagnosis

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SUMMARY

A 43-year-old man presenting with acute coronary syndrome with ST segment elevation myocardial infarction underwent urgent coronary angiography. During the intervention, the patient was found to have several multiple filling defects with dynamic obstruction in the left coronary circulation. Thrombectomy was performed on distal left anterior descending artery and 2nd diagonal artery lesions with balloon angioplasty, which was unsuccessful. Considering the dynamic obstruction in the angiogram, immediate imaging was performed for structural evaluation of the heart. Cardiac CT revealed a circumferential groove on the heart, suggesting an external compression leading to dynamic obstruction of the coronary arteries on angiogram. Cardiac hernia, a very rare aetiology, was suspected to be the culprit for the ST segment elevation myocardial infarction. Thoracoscopy was performed, which revealed congenital cardiac hernia and a fibrous pericardial band encircling the apex. A left mini thoracotomy was performed to release the constriction imposed over the heart, with improvement in circulation.

BACKGROUND

Approximately 90% cases of myocardial infarction are due to acute thrombus. Other causes involve coronary occlusion secondary to vasculitis, valvular disease, emboli, etc. Congenital pericardial defect is a rare clinical entity that may present as angina, palpitation and, rarely, as a catastrophic condition such as acute myocardial infarction, posing a diagnostic dilemma.

Most congenital partial pericardial defect reports have been following incidental findings. These defects usually occur due to premature atrophy of the duct of Cuvier leading to lack of blood supply.¹ Cardiac herniation is often difficult to diagnose at initial presentation and only sporadic reports are available in the literature.^{2 3}

Furthermore, a congenital pericardial defect causing cardiac hernia and presenting as acute myocardial infarction has rarely been reported.^{4 5} We report a rare presentation of an acute ST segment elevation myocardial infarction in a patient following an uncommon event of cardiac herniation secondary to congenital partial pericardial defect.

CASE PRESENTATION

A 43-year-old man presented to the emergency room with sudden onset of chest pain at rest. The pain was graded 10/10; it was a typical chest pain with mild discomfort in breathing and was not relieved with nitroglycerin. The patient had no known significant medical illness and denied smoking, alcohol abuse or illicit drug use, and had no family history of significant medical illness. On physical examination, his vitals were remarkable for elevated blood pressure of 179/94 mm Hg. Physical examination revealed an obese male in severe distress.

INVESTIGATIONS

Initial investigations revealed normal sinus rhythm rate (84) with ST elevation in leads I, II, aVL and V4-V6 (figure 1). Laboratory tests were significant

Figure 1 The initial ECG conducted in the emergency room revealed sinus rhythm, HR: 81 bpm, possible left atrial enlargement, PR interval 166 ms, QRS 96 ms and ST elevation in leads I, aVL, II and V4-V6.

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Figure 2 Coronary angiogram showing dynamic obstruction with two-vessel disease involving distal left anterior descending (LAD) and left circumflex (LCX) arteries. In the distal LAD artery, there was a 99% stenosis. In the first, second and third diagonal arteries, there was a discrete 95% stenosis (see arrows in (A) and (B)). The circumflex vessel was enlarged. There was a 95% stenosis in the distal third of the vessel segment in the first obtuse margin. In the second obtuse margin, there was a 90% stenosis in the distal third of the vessel segment. (A), (B) and (C) during diastole show the prominent filling defect, whereas (D), (E) and (F) show coronary flow. In (C) and (F), the RCA has better flow during both systolic and diastolic phase.

for elevated troponin-I: 11.1 (normal: <0.033 ng/mL), with normal creatinine clearance.

DIFFERENTIAL DIAGNOSIS

Diagnosis of acute ST segment elevation myocardial infarction was made, and the patient was immediately taken to the cardiac catheterisation laboratory.

TREATMENT

Immediate cardiac catheterisation revealed multiple filling defects in the left circulation system. There was two-vessel disease involving the left anterior descending (LAD) artery and left circumflex artery. There was 99% stenosis in the distal LAD. In the first, second and third diagonal arteries, there was 95% stenosis. In the first obtuse margin (OM1) and OM2 there was a 95% and 90% stenosis, respectively. A left ventriculogram revealed overall normal ejection fraction (EF). The immediate impression was that multiple embolic thrombi were occluding these vessels, so percutaneous coronary intervention was attempted on these culprit lesions. Thrombectomy was performed on the distal LAD and second diagonal artery lesions using balloon angioplasty, which was unsuccessful, with 95% residue. The postprocedure impression was a dynamic obstruction of coronary flow secondary to compression of these multiple coronary vessels, from an external source (figure 2). Echocardiography revealed moderate to markedly reduced left ventricular EF. There was akinesis of the mid-apical anterior, apical inferior, apex, distal, mid to distal lateral, and mid to distal posterior walls. Moderately increased thickness of the distal half of the lateral wall was suspected. Doppler parameters were consistent with a reversible restrictive pattern, indicative of decreased left ventricular diastolic compliance and increased left atrial pressure with grade 3 diastolic dysfunction (figure 3).

To confirm the diagnosis, immediate structural evaluation of the heart was performed with imaging studies. CT of the thorax showed a cardiac lesion with increased attenuation, arising from the apex (figure 4). Owing to the dynamic obstruction and suggestive CT imaging finding, a rare condition of cardiac herniation was suspected. MRI further confirmed a constricted apex due to herniation through the existing partial pericardial defect.

The patient was immediately evaluated by the cardiothoracic vascular surgeon. With three-dimensional (3D) CT imaging, structural evaluation of the heart and pericardium was carried out, and a decision to undertake urgent operative intervention was made (figure 5). Images revealed constriction of the coronary arteries and fibrous pericardial rim around the heart (figures 6 and 7). Left thoracoscopy was performed, and an obvious defect in the pericardium was visually identified and herniation of the heart was confirmed.

OUTCOME AND FOLLOW-UP

A left mini thoracotomy was performed; an incision was made on the constricted band of the pericardium all the way to the aortic arch, thus releasing the constriction imposed on the heart. The venous congestion improved significantly and the heart turned pinker in appearance following the release. ST elevation of the ECG resolved during the hospital course and the patient was subsequently discharged with postsurgical follow-up (figure 8).

DISCUSSION

Congenital pericardial defect has generally been classified as either complete or partial. Complete absence of the left pericardium is the most common, accounting for 76% of defects.⁶ Most congenital partial pericardial defect reports have been subsequent to incidental findings. These defects usually occur due to premature atrophy of the duct of Cuvier leading to lack of blood supply.⁶⁻⁷ These small defects may present as angina, palpitation or, rarely, more serious consequences, such as cardiac incarceration.⁷⁻⁸

Difficulty in diagnosis in such catastrophic situations is always a challenge. Physical examination, ECG and chest radiography do not show distinct pathognomonic features. A few case reports have suggested that echocardiography can be the first imaging modality to show unusual findings.⁹⁻¹⁰ Cardiac hypermobility, a 'teardrop' appearance, paradoxical or flat systolic motion of the inter ventricular septum, severe tricuspid regurgitation and right ventricular dilation, are typical for complete absence of pericardium, whereas dilation of the left atrium and

Figure 3 ECHO showed overall moderately to markedly reduced left ventricular function. There was akinesis of the mid-apical anterior, apical inferior, apex, distal septal, mid to distal lateral and mid to distal posterior walls. There was thickening and increased echogenicity of the pericardium of the lateral wall with evidence of less echo dense material between the parietal and visceral pericardium of the distal 1/2 of the lateral wall.

Figure 4 CT of the heart. (A) Coronal and (B) sagittal sections of the thorax showing a circumferential groove on the heart due to external compression caused by the fibrous pericardial rim (see arrows), responsible for dynamic obstruction of the coronary.

Figure 5 Coronal view three-dimensional CT image showing structural evaluation of the heart and constriction by the fibrous pericardium.

regional bulging of the left ventricle is frequently reported in partial absence of the pericardium.⁹

Coronary angiogram might show evidence of dynamic obstruction, as in our case, with no improvement following angiographic intervention, due to the external pressure caused by the pericardial defects on the vessel, secondary to cardiac herniation.

In conclusion, an angiographic intervention finding, such as in this case, with linear circumferential dynamic obstruction, should raise a suspicion of cardiac herniation. The differentials of such a condition would be a left ventricular aneurysm, cardiac tumour, pericardial tumour, thoracic tumour, etc. MRI represents the gold standard test to diagnose this malformation, with the benefits, over CT, of not needing intravenous contrast administration and not subjecting the patient to radiation exposure.⁹ In our case, reconstructed 3D CT imaging was conducted for structural evaluation, which clearly diagnosed this condition.

Thus, imaging should be performed in cases similar to the present case. Intervention should be completed early before

incarcerated hernia develops, in order to prevent irreversible myocardial damage, cardiogenic shock and death.

Learning points

- ▶ In conclusion, an angiographic intervention finding with linear circumferential obstruction, such as in this case, should raise a suspicion of cardiac herniation. The differentials would be a left ventricular aneurysm, cardiac tumour, pericardial tumour, thoracic tumour, etc.
- ▶ MRI is the gold standard test to diagnose this malformation.⁹ But structural evaluation by three-dimensional CT imaging, as in our case, proved obviously diagnostic.
- ▶ Imaging should be performed in cases similar to the present case. Intervention should be completed early before incarcerated hernia develops, in order to prevent irreversible myocardial damage, cardiogenic shock and death.

Figure 6 Sagittal view three-dimensional CT image showing structural evaluation of the heart and compression on the coronary arteries.

Figure 7 Sagittal view three-dimensional CT image showing fibrous rim of the pericardial defect.

Figure 8 Follow-up ECG no longer showing ST elevation, with marked T wave inversions.

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