CASE REPORT

Necrotising coronaritis with fatal outcome

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SUMMARY

A 56-year-old woman presented with acute onset of typical chest pain. She was diagnosed with acute coronary syndrome with ST-segment elevation myocardial infarction. Although significant obstructive coronary artery disease was ruled out by coronary angiography, cardiac MRI showed transmural necrosis of the lateral free wall with extensive microvascular obstruction consistent with ischaemic heart disease. Within 48 hours after initial presentation, the patient suddenly arrested due to pulseless electrical activity with futile resuscitation efforts. Autopsy revealed myocardial perforation with extensive haemothorax due to pericardial laceration, caused by the mechanical chest compressions. Eventually, histology identified diffuse necrotising coronary vasculitis as a rare cause of ischaemic heart disease.

BACKGROUND

Acute chest pain is one of the most frequent reasons for emergency consults. A wide range of cardiovascular, pulmonary, gastrointestinal and musculoskeletal conditions are potential causes. In a recent emergency survey, acute myocardial infarction accounted for 3%–7% of emergency admissions due to acute chest pain.¹ In most cases, atherosclerotic coronary artery disease with partial or complete obstruction is identified as the cause. Here, we present a case with a rare cause of an acute coronary syndrome due to diffuse necrotising coronaritis of unknown aetiology leading to a small transmural myocardial infarction and with fatal myocardial rupture within 48 hours after the initial presentation.

CASE PRESENTATION

A 56-year-old, previously healthy, female patient presented with sudden onset of prolonged acute chest pain to the emergency department with features of acute coronary syndrome with discrete ST-segment elevation in the leads I and aVL and more pronounced ST-segment depression in the inferior leads including V5 and V6 (figure 1). Active smoking and positive family history (father with myocardial infarction) were the only known cardiovascular risk factors.

INVESTIGATIONS

Emergency coronary angiography via a radial approach was performed. There was diffuse atherosclerosis of the epicardial arteries; however, no significant obstructive coronary artery disease or acute vessel occlusion could be identified (figure 2). Monoplane ventriculography revealed a normal systolic function with no wall motion abnormalities.

A clear increase of the cardiac markers was evident, high-sensitivity troponin peaked at 0.72 µg/L ($n < 0.014$) with a maximum rise of creatine kinase to 607 U/L ($n < 192$). D-dimer (< 0.5 mg/L) as well as C-reactive protein (< 5 mg/L) remained within normal ranges.

To further evaluate the aetiology of the cardiomyopathy, cardiac MRI was performed on day 2 after admission on a 1.5 Tesla scanner using a dedicated 32-channel torso coil. Cine imaging with steady state free precession gradient echo revealed overall a mildly reduced systolic function with an ejection fraction of 57% with akinetic myocardium anterolateral at the base of the heart. T2-weighted imaging was performed in the cardiac short-axis direction using dark-blood T2-weighted short-tau inversion recovery fast-spin echo, revealing an elevated signal intensity in these segments consistent with tissue oedema (figure 3). Also, T1-weighted contrast-enhanced inversion-recovery gradient echo was used to assess myocardial delayed enhancement obtained 10 min after contrast injection. The entire anterolateral basal segment extending to the mid-ventricular segment showed transmural delayed contrast enhancement with associated microvascular obstruction (figure 4).

DIFFERENTIAL DIAGNOSIS

Ischaemic heart disease is by far the most common cause of chest pain. In this case, the clinical presentation, the ECG and the increased troponin were consistent with the diagnosis of an acute coronary syndrome. In addition, acute myocarditis was an important differential diagnosis after ruling out significant obstructive coronary artery disease by coronary angiography. However, cardiac MRI revealed microvascular obstruction within the transmural zone of 'late gadolinium enhancement.' Therefore, ischaemic heart disease was obvious as these unique findings are only present in ischaemic heart disease. Transient occlusion of a coronary artery within the distribution territory of the basal anterolateral segment was postulated causing transmural myocardial necrosis. Because no relevant obstructive coronary artery disease or no abrupt vessel occlusion was detected, either spontaneous reperfusion of the coronary artery or severe vasospasm was a possible cause for the myocardial necrosis.

TREATMENT

Initial observation at the intensive care unit was uneventful. The patient was transferred to a regular ward, but maintained under cardiac

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Figure 1 12-lead ECG.

monitoring. Medical treatment included aspirin, ticagrelor and atorvastatin.

FOLLOW-UP AND POSTMORTEM ANALYSIS

On the following day, mild chest pain reappeared. A few hours later, the patient suddenly arrested on the ward with documented pulseless electrical activity. Immediate cardiopulmonary resuscitation using a mechanical chest compression system and repetitive administration of catecholamines was performed; however, no spontaneous circulation could be restored, and reanimation efforts were futile. On fast echo there were no left ventricular contractions and no signs of acute right heart failure; furthermore, no pericardial fluid could be identified.

Postmortem examination revealed a subacute anterolateral myocardial infarction at the base of the heart with transmural myocardial rupture. Almost 3 L of fresh blood was found in the left chest cavity with disrupted pericardium. Most probably, chest compression resulted in pericardial rupture with drainage of the blood into the left chest cavity. Surprisingly, postmortem

Figure 2 Left coronary artery in 90° left lateral projection.

Figure 3 Cardiac magnetic resonance (CMR): Dark-blood T2-weighted short-tau inversion recovery short-axis imaging showing tissue oedema (white asterisk).

analysis revealed a lymphoplasmacytic necrotising coronaritis of all three epicardial vessels with no other signs of a systemic vasculitis (figure 5).

DISCUSSION

This case demonstrates a rare but devastating complication of a comparably small myocardial infarction. Myocardial necrosis

Figure 4 Cardiac magnetic resonance (CMR): T1-weighted 'late enhancement' sequence: short-axis image with transmural 'delayed contrast enhancement' of the basal anterolateral segment (black arrows), lack of signal within the contrast-enhanced myocardium represents extensive microvascular obstruction (white asterisk).

Figure 5 Histological specimen of a coronary artery with lymphoplasmacytic infiltration of the vessel wall.

initiated spontaneous myocardial rupture with the common presentation of a pulseless electrical activity and subsequent death. Free wall cardiac rupture of the left ventricle occurs most frequently in the anterior and lateral wall segments with an overall incidence of 1%–6% in ST-elevation myocardial infarction with an early incidence peak within 24 hours and a later peak between 3 and 5 days after ST-segment elevation myocardial infarction.^{2,3} With the development of new treatment standards of myocardial infarction, in particular the routine use of percutaneous coronary intervention, the incidence of cardiac rupture decreased. However, anterior/lateral infarction, female sex and age over 70 years remain important risk factors for cardiac rupture.⁴ In the modern treatment era, the incidence of free wall cardiac rupture in a recently published series was 1.3% with almost 50% of the patients experiencing rupture within 24 hours and with an extremely high mortality of almost 90%.⁵ Successful restoration of the coronary artery patency after occlusion, either due to mechanical desobliteration or, as it is shown in our case, with spontaneous reperfusion, does not always lead to adequate reperfusion at the microvascular level. This phenomenon known as microvascular obstruction or no reflow phenomenon is significantly associated with adverse ventricular remodelling and overall adverse prognosis.⁶

Coronary arteritis is a rare form of coronary artery disease. Various forms of systemic vasculitis may involve major epicardial coronary arteries; however, isolated involvement of the coronary arteries is rare. In the presented case, no antemortem signs or symptoms of a systemic vasculitis were apparent. Accordingly, no involvement of other arteries in different organs was found

in autopsy. The aetiology of the diffuse coronaritis remained unclear. Histological examination revealed signs of an older necrotic isolated arteritis of the coronary arteries, but not giant cells. These microscopic features and the clinical course would best match with the diagnosis of an isolated coronary polyarteritis nodosa leading to myocardial infarction, potentially induced by a transient occlusion of a coronary artery due to vasospasm with the subsequent fatal outcome.

Learning points

- ▶ Myocardial infarction is most often the result of atherosclerotic coronary artery disease, but other rarer causes include isolated coronary vasculitis.
- ▶ Even in the absence of significant obstructive coronary artery disease on coronary angiogram, transmural myocardial necrosis can evolve from other causes, potentially due to coronary vasospasm.
- ▶ Microvascular obstruction is a unique finding of ischaemic heart disease, which can be revealed by cardiac MRI.
- ▶ Even a small myocardial infarction can cause myocardial perforation with usually fatal outcome.

Contributors TF had the idea for the article, managed the case and is the guarantor for the article. NF submitted the pictures from the pathology and wrote the pathological findings. LS and SS performed the literature search and wrote the article. LS was responsible for the data collection. SS wrote the cardiological findings.

Competing interests None declared.

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