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CASE REPORT
Coronary heart disease

Spontaneous coronary artery dissection masquerading as Takotsubo cardiomyopathy: a case report

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Background Although obstructive coronary artery disease (CAD) is the most likely cause of acute coronary syndromes (ACS), nearly one in 20 individuals with clinical myocardial infarction (MI) do not have obstructive CAD. For many such individuals, presentation of Takotsubo cardiomyopathy frequently mimics presentation of MI, though spontaneous coronary artery dissection (SCAD) is an increasingly recognized aetiology of MI in women.

Case summary This case report describes a woman with chest pain, found to have non-obstructive CAD on angiogram and left ventricular apical dysfunction on echocardiogram raising suspicion for Takotsubo cardiomyopathy. Additional suspicion for SCAD led to coronary CT angiogram (CCTA) which ultimately confirmed this diagnosis.

Discussion Familiarity with a differential diagnosis for non-obstructive CAD is less common than that for obstructive coronary disease. This case emphasizes the clinical features that should raise suspicion for SCAD when Takotsubo is presumed and outlines the clinical utility of CCTA in making this diagnosis when angiography is unrevealing.

Keywords Spontaneous coronary artery dissection • Takotsubo cardiomyopathy • Myocardial infarction • Coronary CT angiogram • Case report

Learning points

- For patients presenting with myocardial infarction with non-obstructive coronary arteries (MINOCA), segmental wall motion abnormalities on echocardiogram should raise suspicion for spontaneous coronary artery dissection (SCAD) over Takotsubo cardiomyopathy.
- When SCAD is suspected and coronary angiography is unrevealing, coronary CT angiogram is able to non-invasively characterize vessel caliber and structure and can uniquely confirm the presence of SCAD.

Introduction

When patients present to our hospitals with chest pain, characteristic risk factors, positive cardiac biomarkers, and/or suggestive electrocardiogram (ECG) changes, we are routinely concerned for acute coronary syndromes (ACS). While obstructive coronary artery disease (CAD) is the expected cause of ACS, nearly one in 20 individuals with clinical myocardial infarction (MI) do not have obstructive CAD.[1] For these patients, the differential diagnosis is less familiar. Takotsubo cardiomyopathy is a diagnosis to consider, which is characterized by ventricular systolic apical ballooning on transthoracic echocardiogram (TTE).

Here, we present a patient who fit this diagnostic pattern—MI with non-obstructive CAD and left ventricular apical dysfunction—but in

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whom additional testing subsequently revealed spontaneous coronary artery dissection (SCAD). We describe the presentation of SCAD initially masquerading as MI from obstructive CAD and then as Takotsubo. We then discuss diagnostic features that can raise clinical suspicion of SCAD when Takotsubo is presumed. We additionally introduce the utility of coronary CT angiogram (CCTA) in making this diagnosis.

Timeline

Setting (hours after arrival)	Events
Emergency Department (1)	Patient with hypertension and hyperlipidaemia presents with central chest pressure and left arm numbness. Electrocardiogram (ECG) reveals anterior ST-segment elevations in V1, V2, and aVR; Troponin-T is elevated to 0.33 ng/mL. With concern for ST-elevation myocardial infarction, she is taken for emergent cardiac catheterization.
Cardiac Catheterization Lab (2)	Coronary angiogram reveals minimal luminal irregularities though coronary arteries are noted to be tortuous.
Cardiac Telemetry Floor (12)	Takotsubo cardiomyopathy is suspected and bedside echocardiogram is done revealing impaired systolic function with regional wall motion abnormalities.
	Regional variation on echocardiogram in conjunction with tortuous coronaries on angiogram raise suspicion for spontaneous coronary artery dissection (SCAD) for which coronary CT angiogram (CCTA) is obtained. CCTA confirms the diagnosis of SCAD.
Post-discharge (48)	Pain does not recur and the patient is discharged home on aspirin and metoprolol for SCAD risk reduction. She is closely followed by cardiology and is enrolled in cardiac rehabilitation.

Case presentation

A 66-year-old woman with hypertension, hyperlipidaemia, migraines, and no known CAD presented to the Emergency Department (ED) with chest pain. She awoke from sleep on the day of presentation with headache and pain in her left jaw that evolved to central chest pressure with left arm numbness. She denied associated nausea or diaphoresis and had never experienced these symptoms. She lived alone in a two-story home, and while she was usually independent in her activities of daily living, on the morning of symptom onset even ambulation of stairs resulted in complete exhaustion.

The patient was proud of her self-care and boasted intake of daily vitamins, a strict organic diet, and minimal stress in her daily life as a retired public school teacher; she had never smoked. She denied recent respiratory infections and had not travelled in the past year. Her family history was negative for premature cardiovascular disease.

Despite a trial of her aspirin analogue, White Willow, persistent symptoms led her to present to the ED. Initial evaluation revealed an anxious woman with stable vital signs (temperature 97°F, heart rate 90 b.p.m., blood pressure 133/81 mmHg, respiratory rate 18/min with an oxygen saturation of 98% on ambient air). Physical examination was unremarkable. Cardiovascular exam revealed a regular rate and rhythm with clear S1 and S2 and no murmurs or gallops. Lungs were clear to auscultation and lower extremities were warm without oedema. Labs were notable for a normal D-dimer 394 ng/mL (<500) and a Troponin-T elevation to 0.33 ng/mL (<0.01). ECG revealed normal sinus rhythm with 1 mm anterior ST-segment elevations in V1, V2, and aVR (*Figure 1*).

Initial treatment in the ED included 325 mg of Aspirin, 80 mg of Atorvastatin, two doses of 0.4 mg of sublingual nitroglycerine, and an unfractionated heparin drip. Nitroglycerine allowed for complete resolution of chest pain but given persistent ECG changes and troponin elevation, P2Y12 inhibition was deferred with management focused on urgent transfer to the cardiac catheterization lab. Angiogram revealed only minimal luminal irregularities though coronary arteries were noted to be tortuous (see Supplementary material online, *Video S1A* and *B*). Takotsubo cardiomyopathy was then considered for which ventriculography was attempted. During catheter insertion, however, the patient sustained transient ventricular fibrillation requiring defibrillation after which further invasive diagnostic imaging was deferred.

To further guide diagnosis, a TTE was obtained. TTE revealed impaired systolic function (ejection fraction of 41%) and regional wall motion abnormalities at the mid anteroseptum, mid inferoseptum, and apical septum but basal wall motion was preserved (see Supplementary material online, *Video S2*). While Takotsubo seemed most likely given the apical wall motion abnormalities with basal sparing and lack of obstructive CAD on angiography, several features remained atypical: (i) history revealed no antecedent stressor, (ii) angiogram revealed coronary tortuosity, (iii) apical wall motion abnormalities on TTE were asymmetric, and (iv) chest pain was not only recrudescent but nitrate-responsive. These features raised suspicion for SCAD not evident on luminal assessment by coronary angiography. A CCTA was then obtained. CCTA not only confirmed absence of calcified coronary artery plaques but additionally noted abrupt caliber changes in the left anterior descending artery (LAD) and its septal branches consistent with SCAD (*Figure 2*).

The patient was started on low dose beta blockade (Metoprolol 50 mg daily) and continued on a low dose Aspirin (81 mg daily). Given her untreated hyperlipidaemia (total cholesterol 296 mg/dL, LDL cholesterol 192 mg/dL), she was additionally discharged on a high dose statin (Atorvastatin 80 mg daily). At 1 month follow-up after discharge, a repeat TTE revealed improvement in her ejection fraction to 78% but with persistent anteroseptal dyskinesis. A repeat CCTA is planned. The patient subsequently completed cardiac rehabilitation and has not had additional medical events.

Discussion

This case illustrates a healthy woman presenting with symptoms consistent with conventional ACS, unrevealing coronary angiography, and presumed Takotsubo by echocardiography, ultimately

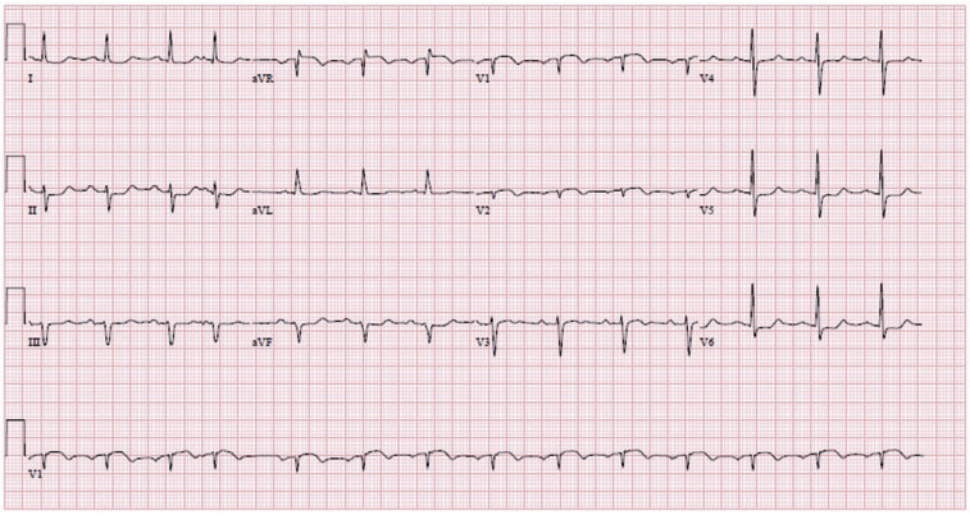

Figure 1 Electrocardiogram on presentation revealing normal sinus rhythm, left axis deviation, and ST-segment elevation in V1, V2, and aVR concerning for ST-elevation myocardial infarction.

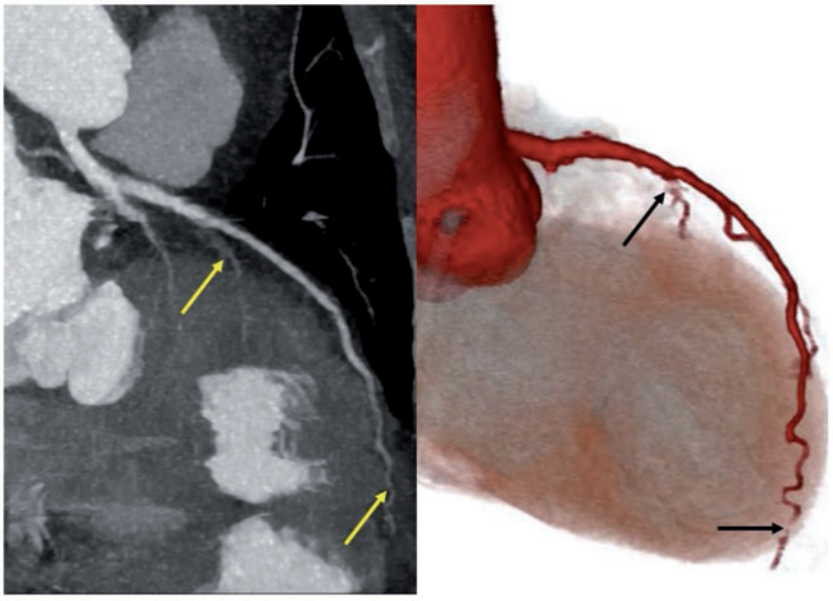

Figure 2 Curved planar formatted (left) and three dimensional volume rendered image (right) of coronary CT angiogram showing abrupt caliber changes in the septal branch (proximal yellow and black arrows) and distal portion of the left anterior descending artery (distal yellow and black arrows).

found to have SCAD by CCTA. The clinical features of ACS that may prompt further non-invasive cardiovascular testing when coronary angiography is unrevealing but suspicion for SCAD is high, are discussed.[2]

SCAD, believed to be intimal tearing or rupture of the vasovasorum leading to bleeding in the arterial wall, is a cause of non-atherosclerotic MI, most typically afflicting younger women.[3] Though the precise cause of arterial wall compromise in these patients is not known, arteriopathies due to fibromuscular dysplasia, connective tissue disorders such as Marfan's and Ehlers–Danlos and high levels of oestrogen and progesterone during pregnancy have all been associated with increased risk.[4]

Given the inability to distinguish coronary plaque rupture from coronary dissection by clinical presentation alone, these patients are appropriately referred for urgent coronary angiography. If coronary artery obstruction is absent in the presence of elevated troponin, the patient's syndrome is consistent with myocardial infarction with non-obstructive coronary arteries (MINOCA) (MI in non-obstructive CAD) for which the differential diagnosis includes Takotsubo, SCAD, coronary vasospasm, coronary embolus, plaque erosion, and myocarditis.[5] Echocardiogram, as emphasized in this case, can provide important clues to direct further evaluation. A recent comparison of Takotsubo and SCAD pointed to the global wall motion abnormalities expected in Takotsubo compared with the focal abnormalities extending along a single epicardial vascular distribution in SCAD (most commonly that of the LAD).[6] As in this case, the septal and apical hypokinesis on TTE and territorial changes on ECG supported the CCTA findings of dissection along the LAD and its septal branches.

When SCAD is evident on angiography, it is radiographically classified into three subtypes.[7] Multiple radiolucent lumens is supportive of Type 1 SCAD, while diffuse stenoses of varying severities and lengths are supportive of Type 2 pathology. Type 3 requires an additional level of suspicion as angiography mimics atherosclerosis and requires intracoronary imaging to make the diagnosis. In this case, SCAD was not seen on angiography but coronary tortuosity, among other atypical features, heightened suspicion of this diagnosis. Given the risks associated with intracoronary imaging, including extension of dissection, CCTA was pursued as a non-invasive way to further characterize vessel caliber and structure. This imaging modality was ultimately able to confirm presence of SCAD and should be considered as the next diagnostic step in patients for whom clinical suspicion remains high despite negative angiography.

Supplementary material

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

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

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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