

Left Main Coronary Artery Spasm Presenting as Non-ST-Elevation Acute Coronary Syndrome in a Patient with Situs Inversus Totalis: A Case Report

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DOI: <https://doi.org/10.36347/sjmcr.2026.v14i10.055>

| Received: 27.08.2026 | Accepted: 10.10.2026 | Published: 28.10.2026

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Abstract

Case Report

Introduction and importance: Situs inversus totalis (SIT) is a rare congenital mirror-image arrangement of the viscera. Coronary artery spasm (CAS) is a transient, intense vasoconstriction of an epicardial coronary artery that can cause acute coronary syndrome (ACS). Their association is exceptional. **Case presentation:** A 51-year-old hypertensive woman presented with prolonged right-sided chest pain. Examination, electrocardiography (ECG), chest radiography and computed tomography revealed previously unknown SIT. The ECG recorded with mirror-image electrode placement showed diffuse ST-segment depression with ST elevation in aVR, and troponin was rising (NSTEMI-ACS). Coronary angiography, performed with mirror-image catheter manipulation, showed a 70–80% left main narrowing reproducing the chest pain, which resolved completely after intracoronary isosorbide dinitrate. Medical treatment led to a favourable outcome. **Clinical discussion:** In SIT, ECG recording and coronary angiography must be adapted to the mirror-image anatomy. Left main spasm must be distinguished from fixed atherosclerotic disease and catheter-induced spasm before any revascularization. **Conclusion:** Intracoronary nitrate testing is mandatory before treating any left main narrowing, particularly in unusual anatomy.

Keywords: *Situs inversus totalis; Dextrocardia; Coronary artery spasm; Left main; Case report.*

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1. INTRODUCTION

Situs inversus totalis (SIT) is a rare developmental anomaly characterized by complete mirror-image transposition of the thoracic and abdominal viscera, with a prevalence of about 1–2 per 10,000 individuals [1,2]. The heart is usually structurally normal, and the prevalence of coronary artery disease is similar to that of the general population [3,4]; however, the reversed anatomy makes the diagnosis and invasive management of ACS challenging [2,5]. Coronary artery spasm (CAS) is a sudden, intense vasoconstriction of an epicardial artery causing total or subtotal occlusion [6], and an established cause of ACS and of myocardial infarction with non-obstructive coronary arteries (MINOCA) [7,8]. Left main involvement is rare but places a large territory at risk [9]. We report spontaneous left main spasm presenting as NSTEMI-ACS in a patient with unknown SIT.

2. CASE PRESENTATION

A 51-year-old woman with recently diagnosed hypertension, treated with an angiotensin receptor

blocker and a calcium channel blocker (CCB), Without other risk factors, presented with prolonged, intense retrosternal pain predominating in the right hemithorax and radiating to the right arm. Blood pressure was 125/75 mmHg and heart rate 68 bpm. Heart sounds were louder over the right hemithorax and the apical impulse was palpated in the right fifth intercostal space. The initial ECG showed decreasing QRS amplitude from V1 to V6 (Figure 1A). Chest radiography showed dextrocardia with a right-sided gastric bubble, and computed tomography confirmed SIT (liver on the left, spleen on the right) (Figure 2). After reversal of the limb leads and right-sided placement of the precordial leads, the ECG showed diffuse ST-segment depression, predominantly anterolateral, with ST-segment elevation in aVR (Figure 1B). Troponin rose from 199 to 578 and then 1,200 ng/L. Echocardiography through right-sided windows showed a non-dilated left ventricle without wall motion abnormality (LVEF 70%).

Citation: Fihri, Y., Faliouni, H., Mouine, N., Benyass, A., & Lakhal, Z. (2026). Left Main Coronary Artery Spasm Presenting as Non-ST-Elevation Acute Coronary Syndrome in a Patient with Situs Inversus Totalis: A Case Report. *Sch J Med Case Rep*, 14(10), 2368-2372. <https://doi.org/10.36347/sjmcr.2026.v14i10.055>

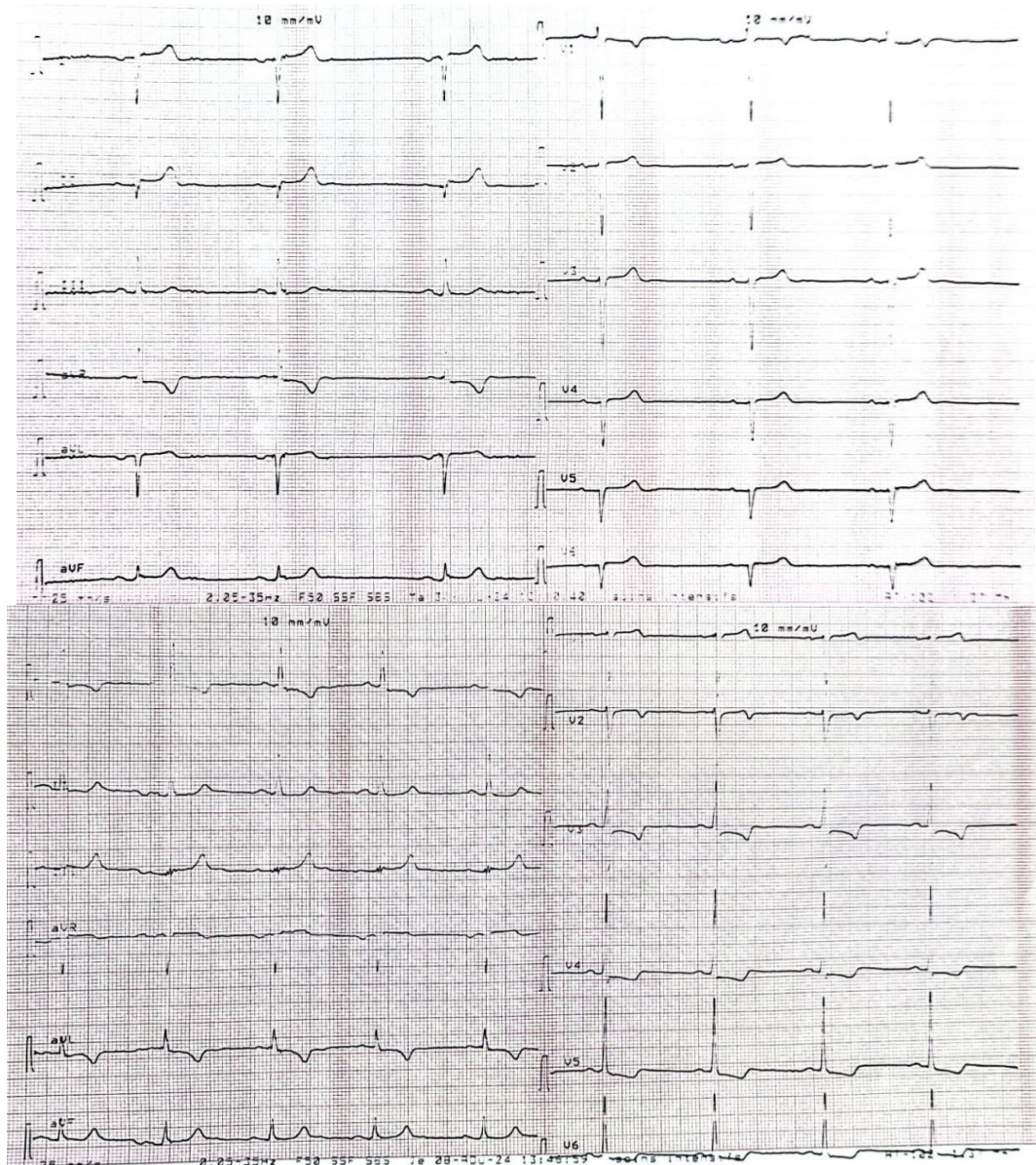


Figure 1: (A) Initial ECG with standard electrode placement: decreasing QRS amplitude from V1 to V6 and apparent ST-segment elevation. (B) ECG after limb-lead reversal and right-sided precordial leads: diffuse ST-segment depression with ST-segment elevation in aVR



Figure 2: (A) Chest radiograph showing dextrocardia and a right-sided gastric air bubble. (B) Abdominal CT showing the liver on the left and the spleen on the right (situs inversus totalis)

The patient underwent invasive coronary angiography [10] via the femoral approach [6 Fr]. Catheters were rotated counterclockwise, the left main was engaged with a Judkins Right 4 (JR4) catheter, and projections were mirrored [11,12]. Angiography showed a tubular 70–80% left main narrowing without other lesions, with recurrence of the usual chest pain (Figure 3A). After intracoronary isosorbide dinitrate, the narrowing resolved completely, with relief of pain and

regression of ECG changes (Figure 3B). The diagnosis of NSTEMI-ACS due to spontaneous left main spasm was made. No revascularization was performed. Treatment comprised dual antiplatelet therapy, long-acting nitrates and a non-dihydropyridine CCB replacing the dihydropyridine; beta-blockers were avoided. The course was uneventful, with no recurrent angina and decreasing troponin with a normal follow-up after 5 months without any angina.

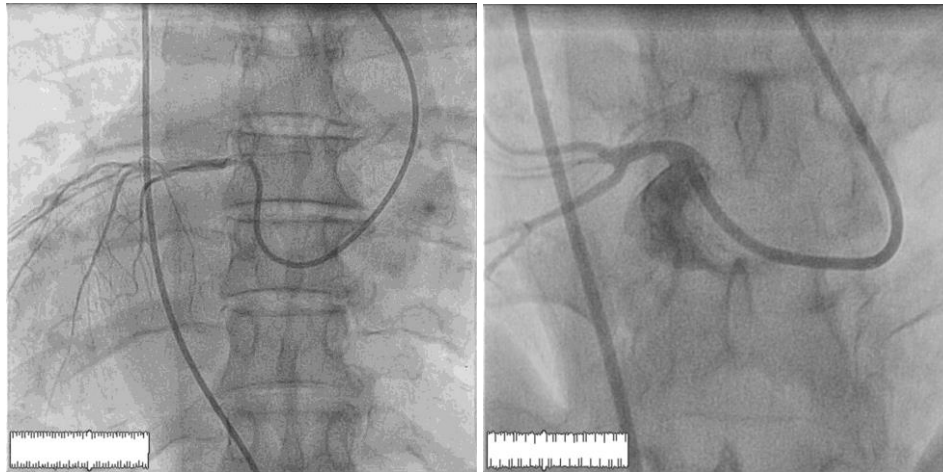


Figure 3: Left coronary angiography (JR4 catheter, mirrored projection). (A) Tight tubular narrowing (70–80%) of the left main with chest pain. (B) Complete resolution after intracoronary isosorbide dinitrate, confirming coronary spasm

3. DISCUSSION

3.1. Situs inversus totalis and dextrocardia

Complete situs inversus with dextrocardia is a rare congenital condition characterized by a mirror-image reversal of the thoracic and abdominal organs, with an estimated prevalence of 1–2 per 10,000 individuals [1,2]. Three main anatomical configurations are classically described: (1) dextrocardia with SIT, in which all organs, including the heart, are mirrored; (2) isolated dextrocardia (dextroversion), with a right-sided heart and normal visceral arrangement (situs solitus); and (3) dextrocardia with situs ambiguus (heterotaxy), frequently associated with asplenia or polysplenia [1,13]. The heart is usually structurally normal in SIT, whereas associated congenital heart defects are frequent in isolated dextrocardia and heterotaxy [1,13]. A proportion of patients with SIT also have primary ciliary dyskinesia (Kartagener syndrome) [1].

Dextrocardia is not an independent risk factor for coronary artery disease [4]. Patients with SIT have a normal life expectancy and an incidence of atherosclerotic coronary disease similar to that of the general population [3,14]. The reversed anatomy may nevertheless delay the diagnosis of ACS and, consequently, timely reperfusion, especially when the anomaly is unknown, as in our patient. Chest pain is often atypical in location—retrosternal or right-sided, radiating to the right shoulder, right arm or jaw—which is thought to reflect transposition of cardiac sensory

afferents to the right-sided upper thoracic segments (T1–T5) during embryological development [5]. A careful physical examination, particularly palpation of the apical impulse in the right fifth intercostal space, and chest radiography (right-sided cardiac silhouette, right-sided aortic arch and gastric bubble) are key to the diagnosis [2].

3.2. Electrocardiographic pitfalls

In dextrocardia with SIT, the standard 12-lead ECG typically shows negative P waves, QRS complexes and T waves in lead I, right-axis deviation, and absent R-wave progression with decreasing QRS amplitude across the precordial leads [2,14]. Unrecognized, this pattern may be mistaken for limb lead reversal or lateral infarction, and may simulate or conceal ST-segment changes. Reversal of the right and left arm electrodes and recording of right-sided precordial leads (V1 and V2 swapped, V3R–V6R) restore an interpretable tracing [2]. In our patient, this manoeuvre changed an apparent ST-elevation pattern into diffuse ST-segment depression with ST-segment elevation in aVR. In the setting of ACS, this pattern of extensive subendocardial ischaemia should raise suspicion of left main or multivessel involvement and, in a patient with ongoing pain, supports an early invasive strategy [10]. Echocardiography also requires adaptation, with the transducer placed in mirror-image positions on the right hemithorax.

3.3. Coronary angiography in situs inversus: technical considerations

Coronary angiography and percutaneous coronary intervention (PCI) are feasible in SIT, but the technique is not standardized and relies on operator experience [5,11]. Because the coronary ostia are mirror images of normal anatomy, catheters must be rotated counterclockwise instead of clockwise. Judkins Right catheters are often better suited to engage the left coronary artery, and Judkins Left or Amplatz catheters the right coronary artery [11]. As the aortic arch is right-sided and the brachiocephalic trunk arises on the left, the left radial approach reproduces the geometry of the usual right radial route, whereas the right radial approach may lead to unfamiliar catheter behaviour. Mirroring the C-arm angulations (left anterior oblique views replacing right anterior oblique views and vice versa), or using the "double-inversion" technique—horizontal inversion of the image combined with reversed oblique angulations—produces images that look familiar to the operator, which facilitates lesion assessment, guidewire manipulation and stent positioning [12]. In our case, the left main ostium was engaged without difficulty using a JR4 catheter, and mirrored projections allowed a reliable assessment of the left main and its bifurcation.

3.4. Coronary artery spasm and left main involvement

Coronary artery spasm, first described by Prinzmetal *et al.*, in 1959 as a "variant form of angina pectoris" [15], results from hyperreactivity of vascular smooth muscle cells, often combined with endothelial dysfunction and autonomic imbalance [16]. It contributes to a broad spectrum of ischaemic syndromes, including stable and unstable angina, acute myocardial infarction, ventricular arrhythmias and sudden cardiac death [16,17]. In the CASPAR study, acetylcholine-induced spasm was found in nearly half of the patients with ACS without a culprit lesion [7]. The COVADIS criteria define definite vasospastic angina by nitrate-responsive angina, transient ischaemic ECG changes and documented coronary spasm—transient total or subtotal (>90%) vasoconstriction with symptoms and ischaemic ECG changes—occurring spontaneously or after a provocative stimulus [6].

Invasive provocative testing with intracoronary acetylcholine or ergonovine remains the reference standard [6,18] and has been shown to be safe and prognostically relevant in patients with MINOCA [19]. Non-invasive methods (ECG during symptoms, Holter monitoring, exercise and hyperventilation testing) are useful but less sensitive [18,20]. In our patient, spasm was documented spontaneously: chest pain, ischaemic ECG changes and a left main narrowing that resolved completely after intracoronary nitrates. Although the visually estimated narrowing (70–80%) was below the threshold used for provocative testing, this sequence strongly supports the diagnosis, and provocation was

considered unnecessary and potentially hazardous given the left main location [9].

From an interventional standpoint, a left main narrowing observed during an acute presentation must be distinguished from two important differential diagnoses. First, catheter-induced spasm, which typically occurs at the catheter tip, is more frequent in the right coronary artery, is usually asymptomatic and is not preceded by ischaemia; in our patient, chest pain, ECG changes and troponin elevation preceded catheterization, which argues for spontaneous spasm. Second, fixed atherosclerotic left main disease: any tubular or ostial left main narrowing should be re-evaluated after intracoronary nitrates before PCI or surgical referral is considered, and intracoronary imaging (IVUS or OCT) is recommended to assess left main lesions and exclude an underlying plaque [21]. Unrecognized left main spasm may otherwise lead to unnecessary stenting or bypass surgery. When the coronary arteries are non-obstructive after nitrates and troponin is elevated, the patient fulfils the definition of MINOCA, and cardiac magnetic resonance is recommended to confirm the ischaemic mechanism and exclude myocarditis or takotsubo syndrome [8,10].

3.5. Management and prognosis

Calcium channel blockers are the cornerstone of treatment, with long-acting nitrates as add-on therapy; beta-blockers, particularly non-selective agents, should be avoided because they may promote spasm, and risk-factor control, especially smoking cessation, is essential [9,21]. Our patient developed spasm despite treatment with a dihydropyridine CCB, which justified switching to a heart-rate-lowering non-dihydropyridine agent combined with nitrates. The benefit of long-term dual antiplatelet therapy in MINOCA due to isolated vasospasm, without evidence of plaque disruption, has not been established, and antiplatelet therapy should be individualized [8,9]. The prognosis of vasospastic angina is generally favourable under calcium channel blockers, but extensive spasm and spasm-related ventricular arrhythmias are associated with a higher risk of adverse events, which justifies close follow-up [9,17].

3.6. Association of situs inversus and coronary spasm

Most published cases of ACS in SIT involve atherosclerotic coronary disease treated with PCI or coronary artery bypass grafting [2,4,5,14]. In an early review, Rosenberg and Rosenberg identified 15 cases of dextrocardia associated with cardiac disease, only 5 of which involved coronary artery disease presenting as angina or infarction-like chest pain [22]. To our knowledge, spontaneous left main spasm in a patient with SIT has not been previously reported. This case illustrates that, beyond the technical adaptations required by the mirror-image anatomy, the usual principles of ACS management apply, and that systematic intracoronary nitrate testing is essential before any

revascularization decision, particularly when the anatomy is unusual.

4. CONCLUSION

SIT should be suspected whenever clinical, ECG or radiological findings suggest an abnormal cardiac position, and ECG and angiographic techniques must be adapted accordingly. Left main spasm in SIT is exceptional; intracoronary nitrates must be given before any left main narrowing is considered for revascularization.

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