### Kazuistika | Case report

# Myocardial infarction in a young patient with antiphospholipid syndrome: A case report

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#### SOUHRN

Tato kazuistika zdůrazňuje význam multimodálního přístupu, personalizovaných léčebných intervencí a nepříznivou prognózu infarktu myokardu bez významného postižení koronárních tepen (myocardial infarction with non-obstructive coronary arteries, MINOCA) u mladého pacienta s antifosfolipidovým syndromem (antiphospholipid syndrome, APS). Na oddělení urgentního příjmu byl přivezen 39letý muž s diagnózou infarktu myokardu s elevacemi úseku ST (STEMI) ve třídě III Killipovy klasifikace. Angiografické vyšetření neprokázalo přítomnost obstrukčního postižení epikardu koronárních tepen, což si vyžádalo další vyšetření srdce magnetickou rezonancí (MR). Toto vyšetření odhalilo jizvy po infarktu myokardu na řadě míst na tepnách a těžkou dysfunkci levé komory. U pacienta došlo k rozvoji srdečního selhání se sníženou ejekční frakcí, a proto mu byl subkutánně implantován kardioverter-defibrilátor.

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#### ABSTRACT

Keywords:
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This case report highlights the importance of a multimodal approach, tailored therapeutic interventions, and the poor prognosis of myocardial infarction with non-obstructive coronary arteries (MINOCA) in a young patient with antiphospholipid syndrome (APS). A 39-year-old man was admitted with a diagnosis of a ST elevation myocardial infarction (STEMI), in Killip class III. Angiography showed absence of obstructive epicardial coronary artery disease prompting further evaluation with cardiac magnetic resonance imaging (MRI). It revealed infarction scars in multiple arterial territories and severe left ventricular dysfunction. The patient evolved with heart failure with reduced ejection fraction and a subcutaneous implantable cardioverter-defibrillator was implanted.

#### Introduction

Antiphospholipid syndrome (APS) is a systemic autoimmune disorder characterized by the presence of antiphospholipid antibodies and a predisposition to arterial and venous thrombotic events.¹ Myocardial infarction (MI) with no obstructive coronary arteries (MINOCA) can be the first presentation of APS.² MINOCA can be defined as: presence of positive cardiac biomarker, clinical evidence of infarction, absence of stenosis (≥50 %) in epicardial coro-

nary arteries on angiography and exclusion of alternative diagnosis.<sup>3</sup> The pathophysiologic mechanisms in MINO-CA are: coronary plaque disruption, coronary vasospasm, spontaneous coronary artery dissection and coronary embolism/thrombosis.<sup>4</sup> The diagnosis of MINOCA requires in addition to coronary angiography and echocardiography, advanced imaging modalities such as coronary intravascular imaging and cardiac cardiac magnetic resonance imaging (CMR) (which is recommended for MINOCA cases where diagnosis is uncertain).<sup>3,4</sup>

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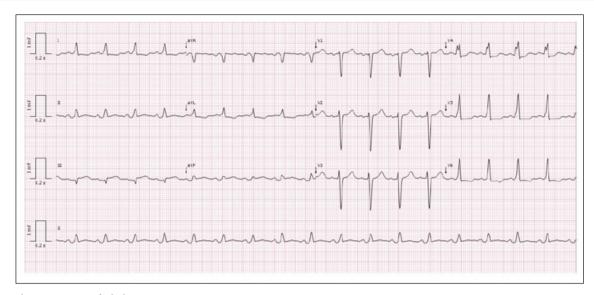


Fig. 1 – EKG on admission.

Concerning prognosis, a meta-analysis revealed an annual mortality rate of 2% in patients with MINOCA.<sup>5</sup> However, a recent study showed that CMR confirmed diagnosis of MINOCA was associated with an increased risk of major adverse cardiovascular events at follow-up.<sup>6</sup>

#### Case report

A 39-year-old man with a history of APS presented to the emergency department with oppressive chest pain and worsening dyspnoea. His medical history included APS with triple positive antibody profile with previous thrombotic events – pulmonary thromboembolism, deep vein thrombosis, arterial ischemia with toe amputation – Raynaud phenomenon, autoimmune haemolytic anaemia, and cryoglobulinemia type 2. Additionally, he had cardiovascular risk factors including arterial hypertension,

dyslipidaemia, and chronic kidney disease secondary to polycystic kidney disease. The patient was on warfarin therapy with labile international normalized ratio (INR) levels in the last months, since recent hospitalizations for infectious causes.

On admission, the patient exhibited tachypnoea, tachycardia, hypoxemia, and hypotension. Pulmonary auscultation revealed diffuse bilateral crackles. Electrocardiogram (EKG) showed sinus rhythm, isolated q waves in DIII with ST segment elevation in DIII, avF and ST segment depression with T wave inversion in  $V_a-V_c$  (Fig. 1).

Laboratory investigation revealed anaemia (Hb 9 mg/dl), worsened chronic kidney disease (creatinine 1.9 mg/dL), elevated troponin T (>10 ng/mL), INR of 4.1, and type 1 respiratory failure. Bedside echocardiogram demonstrated akinesis of the mid-distal segments of the lateral, inferior, and posterior walls, with severe left ventricular (LV) dysfunction.

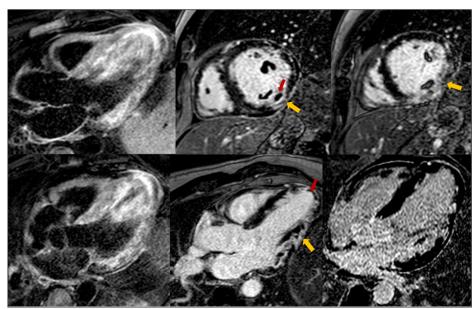


Fig. 2 – Cardiac MRI showing recent inferior and inferolateral infarction (yellow arrows) and intracavitary thrombi (red arrows).

The patient was admitted to coronary care unit with the diagnosis of ST-elevation myocardial infarction (STE-MI) in Killip class III, under intravenous diuretic, non-invasive ventilation and anti-aggregation with aspirin besides his usual warfarin (for INR 3-4). Coronary angiography revealed no obstructive coronary artery disease. Because of the working diagnosis of MINOCA (myocardial infarction with non-obstructive coronary artery disease) he underwent cardiac MRI. The cardiac MRI demonstrated severe LV dilatation and dysfunction, recent inferior and inferolateral infarction and additional non-recent multiple focal subendocardial scars in others arterial territories. We also observed microvascular obstruction at the papillary muscles and intracavitary thrombi in the akinetic regions of inferior and inferolateral walls (the biggest at the distal segment of the inferior wall) (Fig. 2).

Given this, the final diagnosis was recent inferior and inferolateral infarction possibly caused, in clinical context, by coronary thrombosis with spontaneous reperfusion.

Additionally, the patient had previous infarctions in other territories and intracavitary thrombus suggesting embolization/microvascular thrombosis in the setting of APS.

The patient was discharged on a regimen of aspirin, warfarin (target INR 3–4), bisoprolol, lisinopril, empagliflozin, medications targeting associated comorbidities and oriented to cardiology consultation. Medication titration was limited due to hypotension and renal dysfunction and the patient evolved in NYHA II. His INR levels remained controlled. The follow-up echocardiogram demonstrated mild left ventricular dilatation with severe LV disfunction (23%), global hypokinesis and right ventricular dysfunction (TAPSE = 13.1 mm). Given this it was decided to implant a cardioverter-defibrillator. However, in this case, considering the risk of vascular access issues and predisposition to infections, a subcutaneous implantable cardioverter-defibrillator (ICD) was preferred.

#### Discussion

This case report underscores the presentation of MINOCA in a young patient with APS and emphasizes the need for a comprehensive diagnostic approach in these patients. Antiphospholipid syndrome should be considered in young patients presenting with myocardial infarction, particularly in the setting of normal coronary arteries, even when there is no previous history of APS.<sup>1,7</sup> APS is associated with premature atherosclerosis of the coronary arteries, microvascular injury, or coronary thromboembolism, which may trigger myocardial infarction in these patients, 1,8 even in the absence of significant epicardial coronary artery disease. This patient also developed intra-cardiac thrombi, a potentially life-threatening manifestation. Treatment of myocardial infarction in APS is anticoagulation with vitamin-K antagonists (target INR 3.0-4.0). In this patient, aspirin was added because of the recurrent arterial events despite previous anticoagulation and the presence of cardiovascular risk factors. Additionally, concerning the infectious and vascular access issues the ICD implantation was tailored to suit the patient's specific medical situation.

#### Conclusion

Antiphospholipid syndrome can present in the form of venous or arterial thrombosis and should be considered in young patients presenting with myocardial infarction as it can be the first manifestation of the disease. In this MINOCA case, CMR was essential to establish the diagnosis and extension of the myocardial infarction. An ICD is recommended in patients with symptomatic HF of an ischemic etiology and an LVEF  $\leq$  35% despite  $\geq$ 3 months of OMT. In this case, given the high thrombotic risk, we decided subcutaneous ICD implantation, which is a favourable option for patients with vascular access problems or high infectious risk.

#### **Conflict of interest**

The authors declare no competing interests.

#### **Funding**

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#### Ethics approval and informed consent

We declare that the case report has been conducted in accordance with applied ethical standards and guidelines; the Declaration of Helsinki. Appropriate permissions including written informed consent were obtained.

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