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Don't Miss the Sword in the Lung: Unveiling a Previously Undiagnosed Congenital Syndrome in a suspected MINOCA Patient

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Introduction: myocardial infarction with non-obstructive coronary arteries (MINOCA), poses a diagnostic challenge due to its diverse causes. While it is commonly associated with ischemic conditions, there are non-ischemic diseases that can mimic its presentation. As a result, MINOCA is considered a provisional diagnosis until further evaluation confirms ischemia or identifies an alternative cause. Cardiac magnetic resonance (CMR) is an important tool in diagnosing MINOCA and can reveal unexpected findings.

Case history: We report the case of a 57-year-old male with no known cardiovascular risk factors, who regularly participated in sports, presented to the emergency room with several episodes of typical chest pain during exertion and palpitations. On a 12-lead electrocardiogram (ECG), the patient showed subtle, nonspecific ST-segment changes (fig.1) and his high-sensitive troponin-I was elevated (peak value 211 ng/L, normal value 57 ng/L). Upon admission, the patient was asymptomatic and his physical examination was unremarkable: vital parameters, including blood pressure, heart rate, and oxygen saturation, were all within normal limits. Laboratory tests showed normal values for both blood cell counts and C-reactive protein. Transthoracic echocardiography showed normal morphology and function of the left cardiac chambers and excluded valvular and pericardial diseases. However, difficulty in visualizing the right chambers was noted (fig.2). Coronary angiography revealed normal coronary arteries, which raised the suspicion of MINOCA (fig.3).

Fig 1 ECG

Fig 2 Echo

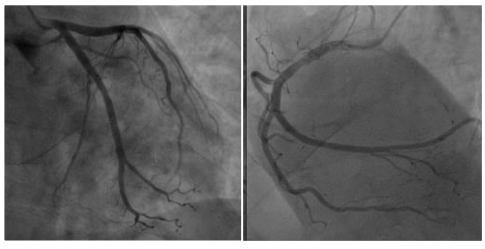

Fig 3 Coronary angiography

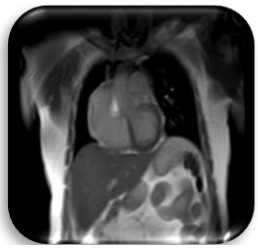

Fig 4 CMR

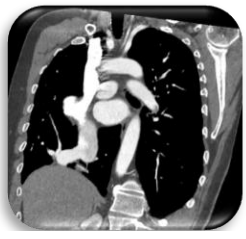

Fig 5 CT scan

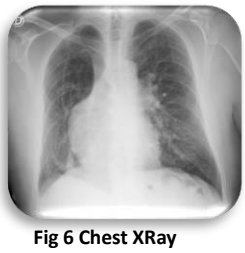

Fig 6 Chest XRay

To investigate the etiology of myocardial damage, the patient underwent cardiac magnetic resonance (CMR) imaging. The CMR showed no evidence of myocardial edema in the T2 mapping sequences and no late gadolinium enhancement (LGE). Nonetheless, CMR uncovered right chamber enlargement (RV EDV 118 ml/m2), along with the ectasia of inferior vena cava (IVC) and pulmonary trunk, attributed to a left-to-right shunt arising from anomalous pulmonary venous return of the right lung in IVC (fig.4). Chest computed tomography (CT) was performed to better define the anatomy of the pulmonary venous return, which revealed a hypoplastic right lung, dextroposition of the heart, right ventricular dilatation and total anomalous venous return of the right lung draining vertically through a large venous collector (the scimitar vein) into the suprahepatic inferior vena cava (fig.5). Abnormal blood vessels from the descending aorta and right inferior diaphragmatic artery were distributed within the right lung base, causing pulmonary sequestration.

A retrospective review of the chest X-ray revealed a reduced right lung area with a partial shift of cardio mediastinal structures to the right and a curvilinear structure extending to the cardiophrenic angle overlying the right heart border (fig.6). All of these findings were consistent with the diagnosis of Scimitar Syndrome. The patient had a favorable outcome during hospitalization, with no recurrence of chest pain. Following discharge, the patient was referred to an adult congenital heart disease center for further evaluation. Surgical intervention was deemed unnecessary due to the lack of symptoms, and regular follow-up appointments were recommended.

Scimitar syndrome is a rare congenital heart defect that is a variant of partial anomalous pulmonary venous return resulting in a left-to-right shunt, with a characteristic anatomical feature resembling a Turkish sword known as 'scimitar.' Adult-onset Scimitar syndrome is a rare condition characterized by symptoms like exertional dyspnea, exercise intolerance, or abnormal heart rhythms. Seeking specialized expertise is crucial for managing this condition in adult patients.

Conclusion: In summary, our case report emphasizes the challenging nature of diagnosing MINOCA and underscores the valuable role of advanced imaging techniques, which can also lead to the diagnosis of previously undetected lifelong cardiac conditions.