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Pulmonary embolism in a patient with a rare coronary anomaly — a clue to the importance of proximal right coronary artery branches

Zatorowość płucna u pacjentki z rzadką anomalią tętnic wieńcowych — wskazówka dotycząca znaczenia proksymalnych gałęzi prawej tętnicy wieńcowej

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Abstract

A 65 year-old female was admitted with acute pulmonary embolism and hypotension. Intravenous streptokinase was administered but severe hypotension persisted. Pulmonary angiography was performed and partial recanalisation of the culprit branch was noted. Echocardiography showed severe right ventricular (RV) dysfunction and free wall akinesia. Due to exaggerated RV dysfunction and severe haemodynamic compromise a coronary angiography was done and showed abnormal origin of right coronary artery (RCA) from left circumflex artery (LCx) with rudimentary RV branches and absent conus artery and right atrial (RA) branch. Inotropic agents were necessary to stabilize the patient's haemodynamic condition for the next four days. We concluded that this coronary anomaly was an important contributory factor in our patient's prolonged unstable condition. This emphasizes the critical role of normal RCA flow under acute haemodynamic stress. Evaluation of the condition of RV perfusion and RCA patency might be justified in any prolonged and disproportionate RV dysfunction following acute pulmonary embolism.

Key words: coronary anomaly, pulmonary embolism, right ventricle

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INTRODUCTION

Right ventricular failure is a critical event in the pathophysiology of pulmonary embolism (PE) and is an important determinant of the patients' outcome in the acute phase [1]. Right ventricular (RV) dilation and hypokinesis as a response to pressure overload may start a vicious cycle of increased myocardial oxygen demand, myocardial ischemia, left ventricular preload reduction, and inability to maintain the cardiac index and systemic pressure, eventually leading to cardiogenic shock [2]. Global RV performance is predominantly determined by the RV free wall (RVFW), which receives blood flow primarily from RV branches of right coronary artery (RCA) [3]. We present a patient with absent proximal RCA branches who developed severe protracted RV dysfunction and haemodynamic compromise following PE.

CASE HISTORY

A 65-year old female, bedridden for over a month due to discopathy, was admitted to our hospital with an acute onset dyspnea and hypotension. She had a history of diabetes and hypertension without any documented history of ischaemic heart disease or chronic lung disease. She had been on valsartan (80 mg twice daily) and glybenclamide (5 mg twice daily). Her vital signs on admission included a blood pressure of 80/60 mm Hg, heart rate of 107/min, respiratory rate of 30/min and O_2 saturation of 82%. She was afebrile. She appeared to be in distress and was covered with cold sweat. Her lungs were clear to auscultation bilaterally and she was tachycardic on cardiac examination. Her routine laboratory tests on admission were within normal limits. Cardiac troponin I (cTnI) level was 2.5 ng/ml (normal = 0.1–1) and D-dimer

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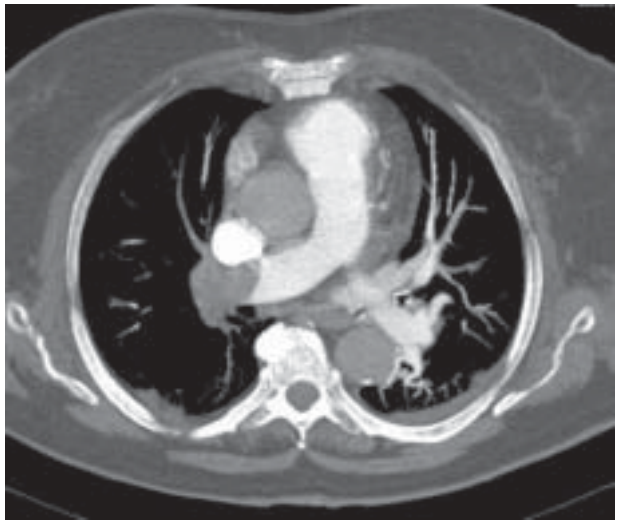

Figure 1. Computed tomography angiography of pulmonary arteries performed immediately after admission. There is subtotal occlusion of the right pulmonary artery (PA) with mild thrombotic involvement in distal part of the left PA branches

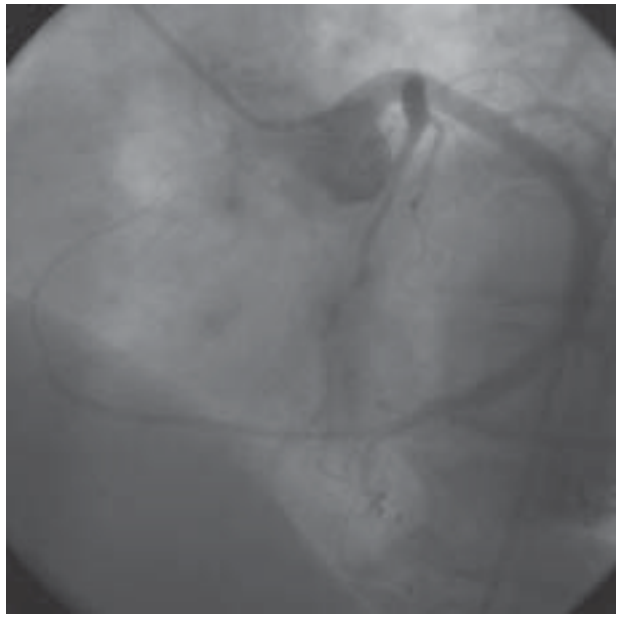

Figure 2. Left anterior oblique view of the coronary angiogram showing the distal portion of the left circumflex artery coursing upwardly to enter the right atrioventricular groove. Note absent conus artery and right atrial branches and rudimentary right ventricular branches

level was 5 mg/L (normal level = less than 0.5). Her electrocardiogram showed sinus tachycardia with an incomplete right bundle branch block pattern and T wave inversion in V1–V4. Bedside echocardiography performed shortly after admission showed D-shape interventricular septum with RV enlargement, severe systolic dysfunction with global RV hypokinesia and free wall akinesia. The patient had normal left ventricular systolic function. A computed tomography (CT) angiogram was done and showed severe thrombotic obstruction of the proximal main right pulmonary artery (Fig. 1) and a small clot in the left lingular lobe artery together with RV enlargement. The patient and her family refused surgical intervention so we decided to proceed with thrombolytic therapy. One and a half million units of streptokinase were administered in 2 hours and she was placed on intravenous infusion of heparin thereafter. No significant improvement was observed in her condition during the following 24 hours so she was scheduled for a pulmonary angiography and probable thrombectomy. The right heart catheterisation pulmonary artery pressure was approximately 42 mm Hg and RV systolic pressure was 48 with an end-diastolic pressure of about 20 mm Hg. At the same time, right pulmonary branch injection showed partially recanalised large thrombus and about 60% diameter stenosis . We considered the possibility of associated RV ischaemia, therefore we proceeded to coronary angiography for evaluation of coronary arteries. Selective left coronary injection showed normal left main, left anterior descending coronary artery (LAD) and left circumflex coronary artery (LCx) but we could not negotiate RCA. Re-evaluation of left coronary artery revealed anomalous origin of RCA from distal LCx (Fig. 2). Considering patient's poor general condition we decided not to proceed with further evaluation. Haemodynamic support of the patient was continued during the following days with inotropic agents. By the fourth day of admission her condition gradually began to stabilize and we could wean her from inotrope agents and she was placed on warfarin. At three months of follow up, she was in stable condition, but her functional class evaluation was not reliable due to her discopathy. Control echocardiographic study showed persistent RV dysfunction despite some improvement in global RV function with only mild improvement of free wall motion index which showed no significant response during dobutamine stress echocardiography. Estimated systolic pulmonary artery pressure with echocardiography was 36 mm Hg. At the same time anomalous origin of RCA from distal LCx was documented in a 64-slice CT angiography of coronary arteries (Fig. 3).

DISCUSSION

Atretic RCA origin and anomalous origin of RCA from distal LCx as its continuation is a very rare condition with less than 20 cases reported in literature. Most published articles emphasize that in the absence of atherosclerotic coronary artery disease this condition is of benign nature with no clinical significance [4]. However the condition of RV seems to be neglected in published literature. Global RV performance is mainly determined by the RVFW, which receives blood flow primarily from RV branches of the RCA [3]. Essential role of RV branches in RV function of patients with inferior wall in-

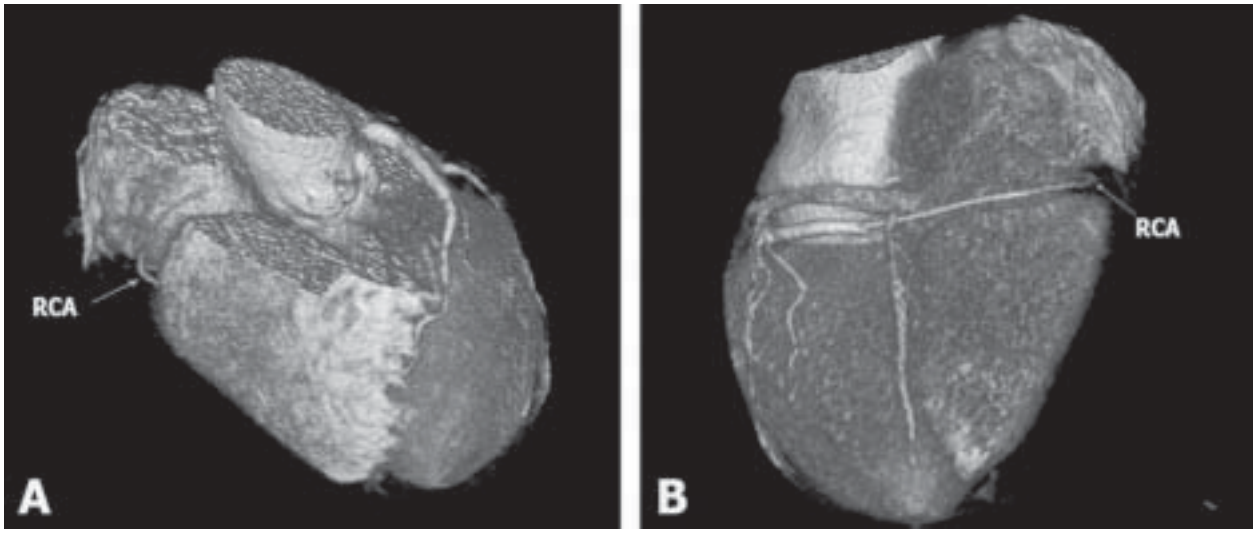

Figure 3. A. Volume rendered multislice computed tomography image in cranial right anterior oblique view showing the absence of right coronary artery (RCA) ostium (arrow); **B.** Diaphragmatic view demonstrating the distal portion of the left circumflex artery coursing upwardly to enter the right atrioventricular groove (large arrow) with no obvious right ventricular branch

farction was shown in a study by Bowers et al in which the status of RV branch perfusion was the critical determinant of RV performance [3]. While normal contraction of RVFW is not necessary in maintaining cardiac function at rest, under right ventricular systolic stress, global cardiac performance becomes increasingly dependent on the RV function. The abrupt rise of the RV afterload with PE leads to increase of wall tension and subsequent chamber dilatation and impaired diastolic and systolic function. Increases in RV wall tension along with decreases in systemic cardiac output and perfusion pressures may disturb the balance between the myocardial oxygen supply and demand, producing ischemia and possibly myocardial infarction [2]. We postulated that the persistent and severe RV dysfunction in our patient might be due to RV hypoperfusion with underdeveloped RCA under a condition of acute haemodynamic stress. Absent conus and right atrial branches together with underdeveloped RV branches showed in the coronary angiography and CT angiogram (Fig. 3), could have also played a role as contributory factors. Lack of significant improvement of RV free wall motion index with dobutamine infusion in our patient, despite mild PAH 3 months after the initial insult, indicates inadequate perfusion of this segment which might have resulted from insufficient coronary perfusion. Underdeveloped right atrial branch impairs right atrial function which is of paramount importance in these settings [5, 6]. This case underscores the role of a normal RCA flow for tolerating the acute RV pressure overload. Further studies are needed to evaluate RV perfusion in patients with anomalous RCA originated from distal LCx.

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