STUDIUM PRZYPADKU / CLINICAL VIGNETTE

Acute respiratory failure due to giant aortic aneurysm mimicking massive pulmonary embolism in a patient with Marfan's syndrome

Ostra niewydolność oddechowa spowodowana olbrzymim tętniakiem aorty imitującym masywną zatorowość płucną u chorego z zespołem Marfana

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A 41-year-old male with Marfan's syndrome was admitted to the emergency department with severe dyspnoea. On his medical history, an operation of tube graft interposition to ascending aorta and aortic valve replacement ten years ago was remarkable. On physical examination, respiratory sound was almost inaudible on the left side of the chest. Cardiac auscultation revealed a grade 2/6 systolic murmur on the left 4^{th} to 6^{th} intercostal spaces. His pulse rate was 108 bpm and blood pressures were measured as 112/73 and 124/82 mm Hg in the left and right arm, respectively. The patient's neck veins were distended. From the analysis of arterial blood gas on 10 L/min oxygen inhalation, pH 7.45, PaCO $_2$ 67 mm Hg, PaO $_2$ 71 mm Hg, SaO $_2$ 80%, and HCO $_3$ ⁻² 20.1 mmol/L were obtained. A chest X-ray revealed widening of the mediastinum and collapsed left lung (Fig. 1). Transthoracic echocardiography revealed mitral valve prolapses, moderate degree of mitral regurgitation, normal left ventricular systolic function (LVEF 60%) and normal functioning of prosthetic aortic valve and significant dilatation and dissection of the descending aorta (Fig. 2). Aortic angiography by computed tomography (CTA) displayed the dissection and dilatation of the descending aorta accompanied by a thrombosed false lumen (Fig. 3A). Dissection membrane was extending from the origin of the left subclavian artery to the right iliac artery and was causing the compression of the left lung and right pulmonary artery (Fig. 3B). The patient was intubated and transferred to the intensive care unit. Unfortunately he died because of acutely developed progressive renal failure, cardiac and pulmonary failure complicated by lung infections over the subsequent ten days.

In patients with Marfan's syndrome, the risk of aortic dissection is approximately 1%. Aortic dissection is a tear within the layers of the aortic wall and the separation of them. An aortic aneurysm may present with symptoms of compression due to mass effect to the adjacent organs. Although the thoracic aorta is a close neighbour to the bronchus, pulmonary atelectasis and obstruction of the pulmonary artery due to an aortic aneurysm is uncommon.

In our case, a giant thoracic aortic aneurysm had reached a large enough size to cause left sided collapse of the lung. Collapsed pulmonary volume and decreased aeration was complicated by oxygen and carbon dioxide exchange and led to hypercapnia and inability to respiratory compensation of acidosis induced by acute renal failure due to aortic dissection. Also, compression of the contralateral right pulmonary artery reduced the perfusion and aggravated the scenario.



Figure 1. Chest X-ray. Widening of the mediastinum and collapsed left lung

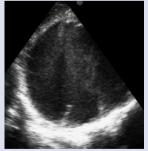


Figure 2. Echocardiography. Significant dilatation and dissection of the descending aorta

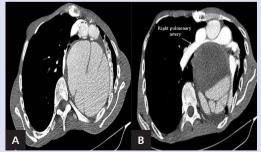


Figure 3. Thoracic CTA; **A**. Dissection and dilatation (11 mm) of the descending aorta with a thrombosed false lumen; **B**. Right pulmonary artery was compressed

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