Unexpected cause of syncope in a young woman

Karolina Supel[1], Michal Zembala[2], Krzysztof Filipiak[2], Jerzy Nozynski[3], Ludomir Stefanczyk[4], Marzenna Zielinska[1]

[1]Intensive Cardiac Therapy Clinic, Medical University of Lodz, Poland
[2]Department of Cardiac Surgery and Transplantology, Silesian Centre for Heart Disease, Katowice, Poland
[3]Department of Histopathology, Silesian Centre for Heart Disease, Katowice, Poland
[4]Department of Radiology, Medical University of Lodz, Poland

Corresponding author:
Karolina Supel MD
Intensive Cardiac Therapy Clinic
Medical University of Lodz
1/3 Sterlinga St
91-425 Lodz, Poland
Phone/fax: +48 42 664 43 64
E-mail:
karolinaewa.k@gmail.com

Submitted: 7 July 2011
Accepted: 12 December 2011

Arch Med Sci 2013; 9, 4: 755–757
DOI: 10.5114/aoms.2013.35224

The cause of syncope in a young woman, previously enjoying good health, turned out to be a great surprise.

An ambulance was called to a 36-year-old woman after she had transiently lost consciousness. The incident was preceded by an intense physical effort, accompanied neither by the irregularity of a cardiac rhythm nor by any chest pain. The patient had not suffered from any disease before.

On admission, the patient was in full verbal and logical contact, without heart failure symptoms or focal neurological signs, with blood pressure of 110/70 mm Hg. During physical examination, there was only a loud holosystolic murmur in the second left intercostal space. Laboratory tests revealed an abnormal N-terminal pro-brain natriuretic peptide (NT-proBNP) value (314.4 ng/ml) and decreased pO_2 (59.5 mm Hg). Troponin T and D-dimer values were normal. Despite regular sinus rhythm 75/min on electrocardiography (ECG), dextrogram and incomplete right bundle branch block were present.

Holter ECG revealed no rhythm and conduction abnormalities. No focal changes were found both in chest X-ray and abdominal ultrasonographic examinations. Computed tomography (CT) of the chest showed a shadow which was interpreted as a massive thrombus with its origin alongside the pulmonary valve and which filled almost the whole right pulmonary artery with preserved peripheral trace flow (Figure 1).

Positron emission tomography (PET) revealed an area of higher radioisotope accumulation in the pulmonary trunk (SUVmax – the maximum standardized uptake value 3.7). Levels of glucose metabolism in other parts of the body were normal.

Cancer markers CA 125, CEA, AFP, CA 19.9, CA 15-3 were normal.

Transthoracic echocardiography showed enlargement of hypertrophic right ventricle and moderate tricuspid valve insufficiency. Right ventricular systolic pressure was estimated at 100 mm Hg. Additional echoes, which receded cyclically in a retrograde manner into the right ventricle outflow tract, showed up in the pulmonary artery lumen. Transesophageal echocardiography revealed a pedunculated tumor growing around the pulmonary valve and leading to complete blockage of the right pulmonary arterial origin. The mass of the tumor filled the pulmonary trunk and right pulmonary branch. A sinuous canal 3–4 mm in diameter appeared inside the tumor mass (Figure 2).

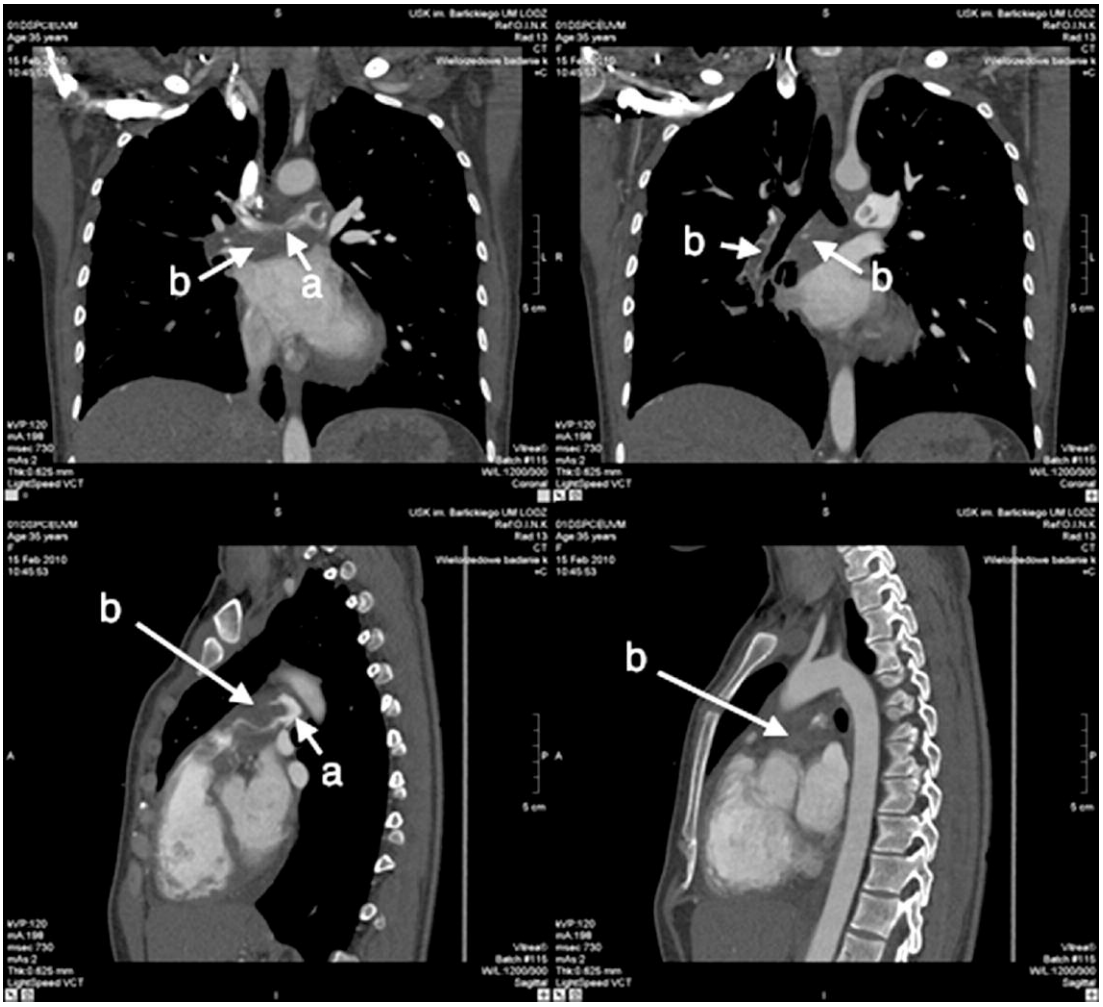

Figure 1. Computed tomography of the chest: (a) residual blood flow to the upper lobe of the right lung, (b) tumor – intimal sarcoma in the trunk and the right pulmonary artery

The decision to perform urgent surgical treatment was made on the basis of gathered examinations. Through median sternotomy, facilitating extracorporeal circulation and moderate systemic hypothermia without cardioplegic arrest the pulmonary trunk was excised. A large, semi-calcified tumor obstructing the right ventricular outflow was visualized, along with a small duct, 5–6 mm in diameter, inside the mass, enabling blood flow to both lungs. The tumor extended from the pulmonary valve, which was also entirely encapsulated within its mass, through the pulmonary trunk to the right and left pulmonary arteries. Successful removal was then carried out, with preservation of the native pulmonary valve. However, removal was incomplete – the tumor mass remained in the lower right lobar artery. The operation and postoperative course were uneventful. Histological and immunohistochemical examination of tumor specimens confirmed the malignant nature of the changes: intimal sarcoma (S-100 protein (–), SMA (+), desmin focally (+), caldesmon (–). Echocardiography, performed 9 days after surgery, showed a residual change of dimensions 17 mm × 13 mm in the pulmonary artery near the wall of the aorta. Chemo- and radiotherapy were scheduled for the patient as a complementary treatment.

Tumors growing out of the pulmonary artery wall in a retrograde manner into the right ventricle lumen occur very seldom. These are primarily undifferentiated sarcomas, leiomyosarcomas or intimal sarcomas. The latter originates from mesenchymal, subepithelial cells, which are components of tunica intima of the great vessels. It occurs twice as often in the pulmonary artery as in the aorta, and rarely in caval veins [1]. A tumor located in the pulmonary trunk often grows peripherally, wallpapering consecutive pulmonary artery branches. In some cases, it bulges towards the right ventricle and causes blockage of the right ventricle outflow tract. Differentiation between leiomyosarcoma and intimal sarcoma is based on immunohistochemical studies. The antigens SMA, desmin, and h-caldesmon occur in leiomyosarcoma, whereas SMA and desmin appear in intimal sarcoma [1]. The average age of patients with intimal sarcoma located

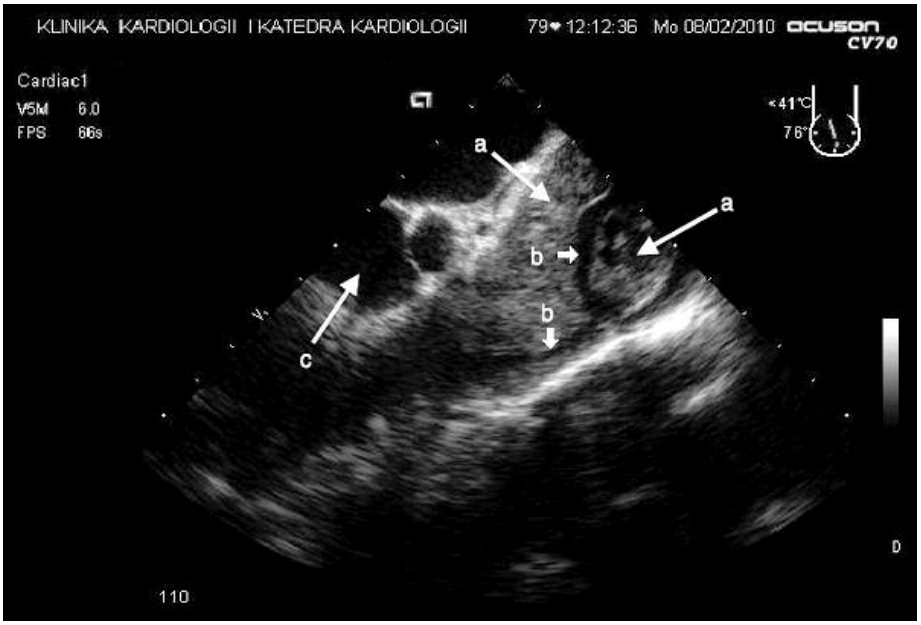

Figure 2. Echocardiogram: (a) tumor – intimal sarcoma in the enlarged pulmonary trunk, (b) residual canal via tumor mass which supplies blood flow to the pulmonary arteries (c) aortic valve

in the pulmonary trunk is 48 years old, and 62 years old for patients in whom it occurs in the aorta, with a higher prevalence in women [2]. Metastases to the lungs occur in 40% of patients. The generalization of cancer with metastases to distant organs (brain, adrenal glands, kidneys) covers 20% of patients [3].

The diagnosis of pulmonary artery neoplasms is established late due to their asymptomatic course and non-specific symptoms. The first symptom of the disease in the presented case was a loss of consciousness. Such a clinical manifestation of intimal sarcoma has not been described before. Significant progression of neoplasm contrasted with lack of overt symptoms of heart failure. Although the tumor mass caused significant flow disturbances in pulmonary circulation, the patient, being a young woman, did not notice any limitations of her exercise tolerance for a long time.

The cause of incorrect diagnosis in cases with vascular sarcomas is the very rare occurrence of such neoplasms. Tumor masses are often misinterpreted as a thrombus and that is why a diagnosis of pulmonary embolism is made. Because such a condition occurs quite often in that age group, a misleading diagnosis of this type is not uncommon. In some cases, the diagnosis of neoplasm is established after biopsy specimen evaluation during surgery or during an autopsy. In the presented case, the tumor mass depicted in CT angiography was similarly interpreted as a thrombus.

Positron emission tomography has raised some hopes of distinguishing these two disorders. According to Ito *et al.* [4], the SUVmax parameter value could be useful in differentiation between pulmonary embolism and pulmonary artery sarcoma. The average SUVmax value of thrombus was 2.31 ±0.41 (n = 10), while for neoplasm it was 7.63 ±2.21 (n = 3). These observations are confirmed by the result of PET examination in the patient. The intimal sarcoma SUVmax value was 3.7, which is higher than values recorded for thrombus.

Intimal sarcoma is characterized by an extremely poor prognosis. The median survival rate after radical surgery is 23.5–36.5 months [5], and after non-radical surgery 11 months [6]. The role of chemo- and radiotherapy as a complementary treatment is uncertain and requires further analysis.

References

1. Donald M. S. Pulmonary and thoracic sarcomas. Curr Diag Pathol 2006; 12: 409-17.
2. Ozbek C, Emrecan B, Calli AO, Gurbuz A. Intimal sarcoma of the pulmonary artery with retrograde extension into the pulmonic valve and right ventricle. Tex Heart Inst J 2007; 34: 119-21.
3. Hirose T, Ishikawa N, Hamada K, et al. A case of intimal sarcoma of the pulmonary artery treated with chemoradiotherapy. Inter Med 2009; 48: 245-9.
4. Ito K, Kubota K, Morooka M, et al. Diagnostic usefulness of 18F-FDG PET/CT in the differentiation of pulmonary artery sarcoma and pulmonary embolism. Ann Nucl Med 2009; 23: 671-6.
5. Bakaeen F, Jaroszewski D, Rice D, et al. Outcomes after surgical resection of cardiac sarcoma in the multimodality treatment era. J Thorac Cardiovasc Surg 2009; 137: 1454-60.
6. Blackmon S, Rice D, Correa A, et al. Management of primary pulmonary artery sarcomas. Ann Thorac Surg 2009; 87: 977-84.