Cardiac papillary fibroelastoma mimicking infective endocarditis

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A 61-year-old woman with a history of hypertension and asthma was transferred to the Institute of Cardiology in Warsaw from another hospital due to a suspicion of pulmonary valve infective endocarditis (IE). The symptoms began five months previously, with progressive weakness, fever up to 39°C, muscle pain, and weight loss of 6 kg. Prior to admission she had been repeatedly hospitalised for over two months. During the first hospitalisation transthoracic echocardiography (TTE) revealed a round mobile echogenic mass attached to the arterial side of the pulmonary valve, without valvular dysfunction. The pulmonary trunk was not expanded. Computed tomography (CT) confirmed the presence of a mobile mass attached to the pulmonary valve. Moreover, diffuse consolidation in both lungs was present. Repeated blood cultures remained negative. Doppler ultrasonography excluded deep vein thrombosis. Abdominal ultrasonography, oesophagogastroduodenoscopy, and gynaecological examination did not reveal any abnormalities. Bronchoscopy demonstrated features of chronic bronchitis. Tuberculosis was excluded. Finally, the patient was diagnosed with IE, and antibiotic therapy was initiated. The treatment with ceftriaxone, vancomycin, levofloxacin, ciprofloxacin, and fluconazole did not improve the patient's status, and she was referred to our centre. On admission, the woman was in good general condition, her pulse rate was 61 bpm and blood pressure was 111/73 mmHg. Laboratory tests showed an elevated level of C-reactive protein (0.07 g/L; reference range, 0.0–0.005 g/L) and a reduced haemoglobin level (90 g/L; reference range, 120-160 g/L). During hospitalisation, fever up to 39.2°C was observed. Blood cultures in our hospital were also negative. TTE confirmed a round mobile additional mass attached to the arterial side of the pulmonary valve (Fig. 1A, B), suspicious of a tumour. Electrocardiogram-gated cardiac CT showed a hypodense, microlobulated mass, sized 16 × 14 mm, attached to the leaflet of the pulmonary valve by a narrow stalk (Fig. 2A, B). The attenuation value was 150 Hounsfield units after contrast administration. Due to morphological features on CT and the location of the lesion, cardiac tumour was suspected. Pulmonary embolism and infection were excluded. The patient was referred for a surgical intervention and underwent a successful removal of the pulmonary valve mass. Histopathological examination showed cardiac papillary fibroelastoma (CPF) (Suppl. Fig. 1 - see journal website). The patient was discharged home in good condition. CPFs are benign primary cardiac tumours. They are the second most common primary



Figure 1. **A**, **B**. Transthoracic echocardiogram. Parasternal short-axis view showing an additional mass within the main pulmonary artery attached to the pulmonary valve (arrows)



Figure 2. Electrocardiogram-gated cardiac computed tomography; sagittal (**A**) and virtual angioscopy (**B**) views showing a lobulated mass attached to the pulmonary valve (arrows)

cardiac tumours, and the most common cardiac valvular tumours [1, 2]. More than 95% of CPFs arise in the left heart [2], whereas pulmonary valve papillary fibroelastomas are extremely rare. CPFs are mostly valvular, < 25% of them arise from nonvalvular endocardial surfaces [2, 3]. The most common location of CPFs is the aortic valve (44%-59%), followed by the mitral valve (13%-35%), the tricuspid valve (4%-15%), and the pulmonary valve (2%-8%) [2, 3]. Predominantly, CPFs are asymptomatic and constitute incidental findings on routine echocardiography, but they may cause life-threatening complications such as stroke, unstable angina, myocardial infarction, sudden cardiac arrest, heart failure, and exceptionally — pulmonary embolism [2]. The most common clinical presentations are caused by embolism and depend on many factors, including tumour location, size, and growth rate. Embolisation may occur from the fragile papillary fronds of the tumour itself or from a thrombus formed on the tumour [3]. Rare cases of CPF manifested with fever, which abated after successful excision of the tumour, were also described [3]. Our case shows difficulty in proper recognition of a cardiac tumour whose clinical symptoms may suggest IE.

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Conflict of interest: none declared

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