

Quadricuspid pulmonary valve complicated with aneurysm of pulmonary trunk diagnosed with ECG-gated computed tomography

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[Received 16 July 2009; Accepted 10 October 2009]

Quadricuspid pulmonary valve (QPV) is an uncommon congenital defect reported in the general population with a frequency of up to 0.25%. The defect usually does not cause severe clinical complications and its presence frequently remains clinically silent. Moreover, there are several difficulties in visualization of pulmonary valve using basic diagnostic modalities such as echocardiography. Therefore, in the majority of cases, QPV is detected accidentally during cardiac procedures or post mortem.

The authors present a case of QPV complicated with aneurysm of the pulmonary trunk, diagnosed with computed tomography in 70-year-old woman. Although the patient had undergone transthoracic echocardiography examinations several times in the past, only computed tomography allowed the detection of the anomalous valve. In addition, the examination confirmed aneurysm of the pulmonary trunk.

To the best of our knowledge, this is the first case of QPV diagnosed in vivo with computed tomography. (Folia Morphol 2009; 68, 4: 290–293)

Key words: quadricuspid pulmonary valve, aneurysm of pulmonary trunk, computed tomography

INTRODUCTION

The quadricuspid pulmonary valve (QPV) is an uncommon congenital defect resulting from inappropriate development of truncus arteriosus and disturbances in the division of the outflow tract in the early stages of embryogenesis [2, 5]. The frequency of the defect is reported to range from 0.02% to 0.25% of the general population [2, 4].

Although QPV frequently appears to be an isolated malformation [6], some authors postulate its association with other congenital cardiovascular anomalies, such as: patent arterial duct, atrial or ventricular septal defects, and abnormalities of the

aortic valve [2, 4, 5, 7]. According to the literature, QPV may also coexist with less common congenital syndromes — transposition of the great arteries [10], interrupted aortic arch with aortic valve atresia [12], or misalignment of pulmonary vessels with alveolar capillary dysplasia [11].

In contrast to the four-leaflet variant of aortic valve, QPV usually does not cause severe clinical consequences; however, there are single reports of the defect complicated with stenosis, insufficiency of the valve, or dilated pulmonary trunk [7, 8]. Consequently, its presence frequently remains clinically silent. and in the majority of cases, the defect is detected

accidentally during cardiac procedures or post mortem [1].

Despite this, of the more than 200 cases of QPV that have been reported until now, only a few of them have been diagnosed *in vivo* using echocardiography or magnetic resonance imaging [4, 5, 8]. To the best of our knowledge, we are presenting the first case of QPV complicated with aneurysm of the pulmonary trunk diagnosed with electrocardiography-gated multislice computed tomography (ECG-MSCT).

CASE REPORT

A seventy-year-old female was admitted to our institution because of unspecific chest pain. The patient was referred to the cardiology unit due to a history of previous treatment. In 1995 she was treated at the pulmonology unit because of haemoptysis. Candida infection was diagnosed and a suspicion of widening of the pulmonary trunk was established on the basis of a chest X-ray. In 1998 the patient underwent myocardial infarct, and since then she had remained under regular follow-up at the cardiology outpatient clinic. She had transthoracic echocardiography (TTE) performed, which confirmed the widening of the pulmonary trunk and, in addition, revealed slight stenosis and regurgitation of the pulmonary valve as well as hypokinesis of the inferior wall of the heart. Echocardiographical findings remained constant in consecutive examinations performed 5 times in one-year intervals. The pulmonary valve was not visualized with regard to its morphology in any of these examinations.

Current clinical examination revealed decreased effort tolerance (NYHA functional class II). Blood pressure was 130/85 mm Hg. ECG, except the signs of past myocardial infarct of inferior wall, was normal (sinus rhythm, heart rate 70/min, normal electrical axis of the heart). The present echocardiographical picture was stable except for a slight insufficiency in both atrioventricular valves. However, the pulmonary valve was not visualised directly.

Because of the clinical history of the patient, the decision to perform ECG-MSCT was made. The examination was carried out in standard cardiac protocol with a 64-row scanner. A bolus of 120 mL of non-ionic contrast medium was administered intravenously through an automatic syringe at 4 mL/s. Multiplanar and 3D reconstructions were acquired using a dedicated workstation. Both coronary arteries and the morphology of the heart were evaluated. The aortic and pulmonary valves were assessed

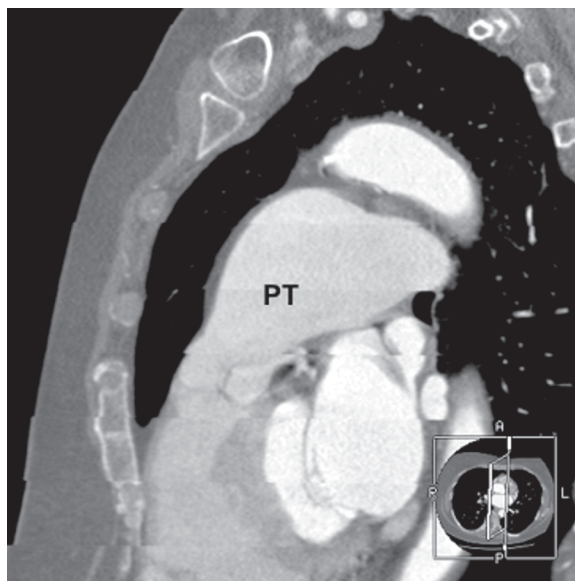


Figure 1. Sagittal view of widened pulmonary trunk (PT) in multiplanar reconstruction.

in every 10% of the ECG R-R segment using oblique reconstructions in the planes parallel to the planes of the valves.

No signs of significant stenosis in the coronary arteries were found. Parietal atheromatous changes were revealed in the left anterior descending artery. The heart was enlarged within the limits of the right chamber — the functional parameters were: end-diastolic volume 182 mL, end-systolic volume 74 mL, stroke volume 108 mL, and ejection fraction 59%. The examination confirmed either hypokinesis of the inferior wall of the heart or aneurysm of the pulmonary trunk, which was dilated in the middle and distal sections to 49 mm in diameter (Fig. 1). Moreover, the anomaly of the pulmonary orifice in the form of a quadricuspid, slightly insufficient pulmonary valve was revealed (Fig. 2, 3). The accessory leaflet was situated backmost and to the right (Fig. 2). Both the accessory cusp and the right one were smaller, approximately equal in size, and partially merged. The remaining two cusps, the anterior and the left one, were larger and were comparable in size to each other. No other malformations of the heart were detected.

To verify the diagnosis of pulmonary valve abnormality, a TTE examination was performed. Again, it was impossible to visualize the pulmonary valve directly, and in general the echocardiographical picture was stable. The pressure in the pulmonary artery was evaluated using the acceleration time method and it was slightly elevated, at up to 40 mm Hg.

Due to the age of the patient and either a stable clinical condition or echocardiographical and radio-



Figure 2. Multiplanar reconstruction presenting quadricuspid pulmonary valve in the projection parallel to the plane of the valve. The arrow indicates the accessory cusp.

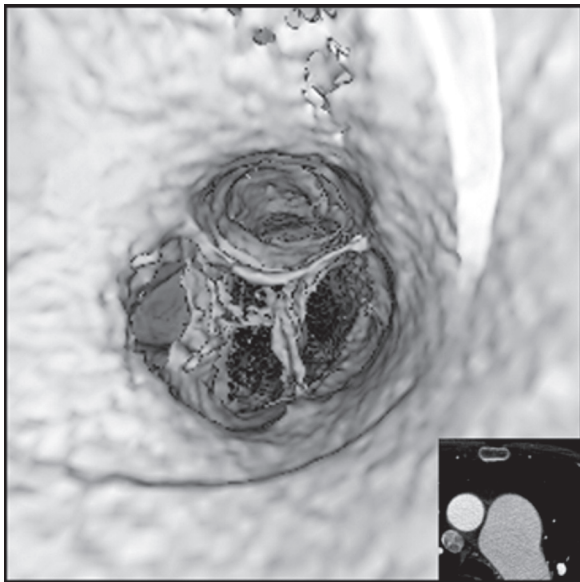


Figure 3. Quadricuspid pulmonary valve in volume rendering reconstruction.

logical picture, conservative therapy in the form of pharmacological treatment was applied. The patient was discharged from the hospital in a good general state, with recommendation of periodic controls in a cardiological out-patient clinic.

DISCUSSION

For many years, the majority of publications have reported QPV detected accidentally during cardiac

surgery or in postmortem examinations; however, nowadays the defect may also be identified with imaging methods such as computed tomography. The accuracy of evaluation of the morphology of pulmonary orifices in computed tomography and the possibility of identification of leaflets and their commissures is obviously related to the technique applied. In earlier studies, because of thicker reconstruction layers (5–7 mm), the precise evaluation of the pulmonary valve was impossible. The introduction of multislice computed tomography with 0.6 mm collimation, quick table feed, and ECG-gating resulted in dynamic development of imaging of the heart's structures and vessels. At present, ECG-MSCT is becoming a widely used imaging modality in patients with suspected heart disease, including valvular pathologies [3, 9]. Its increasing availability allows an *in vivo* diagnosis of rare cardiac anomalies and their clinical complications.

Isolated QPV is usually asymptomatic. In rare cases, symptoms such as fatigue or palpitation of the heart may appear; however, they are not specific and are usually related to valve incompetence or stenosis. Although the malformation does not cause severe clinical complications, the presence of the defect may be important in some specific situations, e.g. requiring Ross procedure [1]. In such cases, preoperative detection of the abnormal valve allows modification of treatment. Therefore, the importance of modern imaging techniques which allow the evaluation of the morphology of pulmonary valves increases.

In the presented case, the patient had undergone TTE several times, but the defect had not been detected in any of these examinations — probably because of difficulties in visualization of the pulmonary valve, which is typical for TTE, and result from its anatomical features and spatial location [8]. In the described case, only ECG-MSCT allowed detailed visualization of the pulmonary valve giving the opportunity to diagnose the less frequent variant of QPV, reported in only 15% of cases of the defect, with two equal and two smaller cusps [4]. Therefore, it enabled explanation of the reason for the pulmonary trunk's dilatation.

CONCLUSIONS

ECG-MSCT may be useful in the evaluation of pulmonary valve morphology, its quadricuspid variant, and possible complications of the defect, as long as radiologists, as well as the assessment of coronary arteries, bear in mind the complex analysis of the heart's structures.

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