



CT Findings of Quadricuspid Pulmonary Valve with Pulmonary Artery Aneurysm

사첨폐동맥판막과 폐동맥동맥류의 컴퓨터단층촬영 영상소견

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Quadricuspid pulmonary valve (QPV) with pulmonary artery aneurysm is an uncommon condition. QPV is typically clinically silent, so it is often diagnosed after death. However, recent advancements in imaging modalities, such as computed tomography (CT), have allowed more frequent incidental diagnosis of QPV in living adults. We report a case of QPV with pulmonary artery aneurysm in an asymptomatic adult; the condition was detected by CT aortography but was not discernible in echocardiography. Following the case presentation, we review the prior related literature.

Index terms

Pulmonary Valve
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INTRODUCTION

Quadricuspid pulmonary valve (QPV) is an uncommon congenital heart morphologic variation. QPV with pulmonary artery aneurysm makes it more of a rare case. QPV usually tends to be clinically silent. Thus, it has mostly been diagnosed post mortem (1). Recently this case has been incidentally found in living patients due to advances in imaging technologies such as electrocardiography (ECG)-gated computed tomography (CT) and magnetic resonance imaging (MRI) (2). However, it is still difficult to visualize QPV in echocardiography (3).

We report a case of QPV with pulmonary artery aneurysm in an asymptomatic adult diagnosed by ECG-gated thoracic aorta CT, however, undetected in echocardiography and review the previous literature regarding QPV.

CASE REPORT

A 58-year-old female patient visited our hospital with an ab-

normal chest radiograph performed at a local clinic. She was a non-smoker and had no respiratory or cardiac symptoms. She had no notable past medical history or family history. Physical examination revealed continuous bruit along the left upper sternal border. Normal sinus rhythm was observed on resting ECG. Chest radiography revealed a bulging opacity in the left hilum, probably due to a prominent vascular shadow, and no other abnormal findings were found. Transthoracic echocardiography (TTE) showed a dilated pulmonary arteries [main pulmonary artery (MPA) = 51.9 mm, left PA = 32.6 mm, right PA = 20.1 mm in diameter] and a Doppler study on parasternal short axis view revealed continuous shunt flow to the MPA, suggesting patent ductus arteriosus (PDA). However, transesophageal echocardiography (TEE) revealed no definite shunt flow to the MPA, indicating a low possibility of PDA. Therefore, retrospective ECG-gated thoracic aorta CT was performed on a 64 detector-row CT scanner (Brilliance 64; Philips Medical Systems, Cleveland, OH, USA). Imaging parameters were as follows: 120 kV, 800–1000 mAs/slice, 64 × 0.625 mm detector col-

limation. Dilated pulmonary arteries (MPA = 43 mm, left PA = 27 mm, right PA = 24 mm) were observed on thoracic aorta CT as well, but it also showed 4 equally-sized pulmonary cusps, corresponding to a QPV. However, her tricuspid aortic valve (AV) was normal (Fig. 1). The patient had no symptoms, so she was put on regular follow up.

DISCUSSION

QPV is an uncommon congenital morphologic variation, usually consisting of 3 equally-sized cusps and 1 smaller one (1). Although it tends to be an isolated malformation, it can be associated with other cardiac anomalies such as PDA, atrioventricular septal defect, and coarctation of aorta (1). However, the most

commonly associated congenital anomaly is AV malformation such as a bicuspid AV. This is due to the common embryogenic pathway of the PV and the AV (4). Both tricuspid valves are normally developed from mesenchymal proliferation in the common trunk and deviation of the aortopulmonary septum during the fourth week of gestational age. Abnormal processes in either mesenchymal proliferation or septal deviation could lead to the development of QPV with an abnormal AV (4). In our case, the patient had a QPV with four same sized cusps and a normal tricuspid AV. This can be better explained by an abnormal process in mesenchymal proliferation rather than septal deviation (1) (Fig. 2). QPV tends to be an isolated malformation and usually remains clinically silent. Because of its benign features, diagnosis of QPV has been usually made after death.

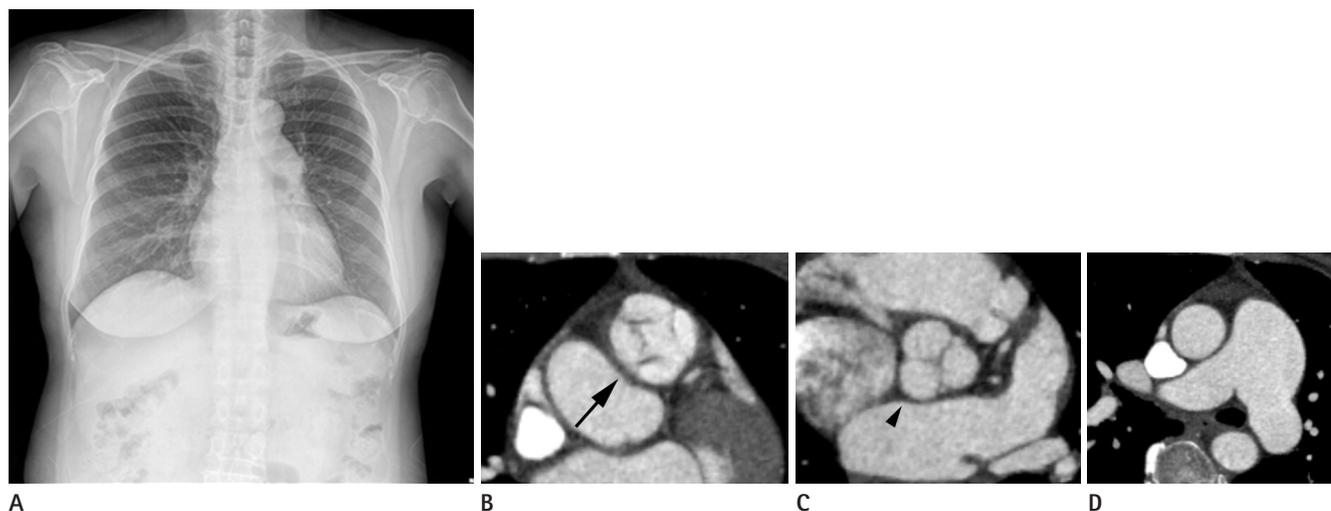


Fig. 1. These figures show QPV with pulmonary artery aneurysm. **A.** Chest radiography shows bulging opacity in the left hilum, probably due to prominent vascular shadow. **B, C.** Double oblique reconstruction CT image shows QPV (arrow, **B**) and normal tricuspid AV (arrowhead, **C**). **D.** Axial CT image shows dilated pulmonary artery, suggesting pulmonary artery aneurysm. AV = aortic valve, QPV = quadricuspid pulmonary valve

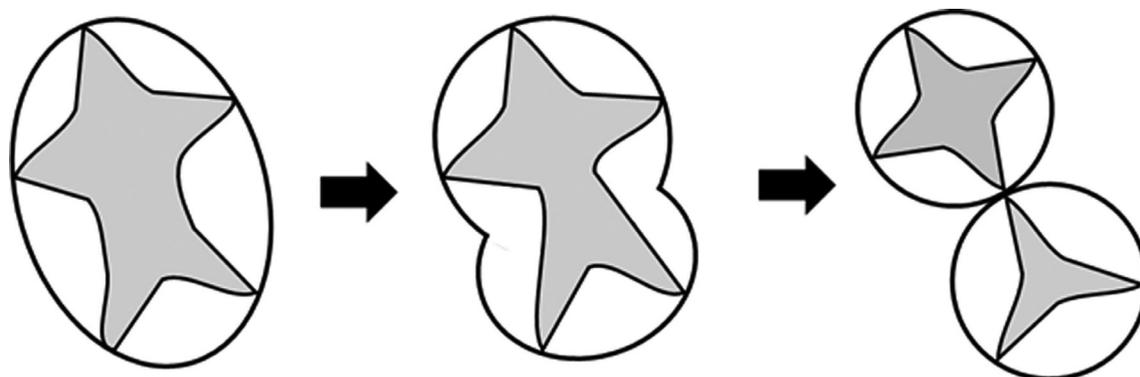


Fig. 2. This figure shows embryogenesis of QPV and normal tricuspid AV. Abnormal mesenchymal proliferation is the first step leading to QPV. AV = aortic valve, QPV = quadricuspid pulmonary valve

According to previous autopsy studies, the frequency of QPV is reported to be from 1/2000 to 1/400 (1). Recent advances in imaging technologies and frequent health examinations have resulted in more incidental diagnosis of QPV. According to a previous report, the prevalence of QPV is reported to be 0.2% in the general population (5). It is predominant in males with a male to female ratio of 2:1 (1).

Pulmonary artery aneurysm is also a rare condition, which is found in about 1/14000 autopsies with no gender predominance (6). There are many causes of pulmonary artery aneurysm. Among them, congenital heart disease such as PDA, atrial septal defect and PV stenosis accounts for 50% of the pulmonary artery aneurysms (7). Only a few cases have no identified cause. To our knowledge, there have only been three previously reported cases of QPV accompanied by pulmonary artery aneurysm. In two cases, the aneurysm was found in the MPA or left PA, while the other case had the aneurysm in both MPA and left PA (2, 8, 9). However, in our patient, MPA, left PA and right PA all showed aneurysmal dilatation. Our case had no notable findings that can be considered as the cause of pulmonary artery aneurysm except the presence of QPV. Therefore, it is probable that the QPV has had some effect on the development of aneurysm. A geometric change of the valve opening area in QPV may increase the pressure, leading to aneurysm development. However, because diameter of the right MPA is smaller than that of the left pulmonary artery, the possibility of mild pulmonary valvular stenosis as a cause of aneurysmal dilatation of the proximal pulmonary arteries is not reliably excluded in this case.

Diagnosis of QPV in a live patient is dependent on imaging studies such as echocardiography, CT, and MRI (2, 3). All studies may be used to visualize the quadricuspid pulmonary cusps. However, diagnosis of QPV by TTE is difficult due to the anatomical disposition of the PV relevant to the thoracic wall (3). Thus, even a skilled physician could miss this anomaly. Although TEE is more accurate than the TTE, TEE may not detect the QPV as in our case. On the other hand, ECG-gated CT and MRI are the most sensitive methods to diagnose QPV. These image modalities not only allow visualization of morphologic features of the valve but also reveal associated structural deformities. According to the Korean guideline for cardiac CT, car-

diac CT is recommended when valvular disease is suspected, and other noninvasive test methods are not considered appropriate (10). Inadequate diagnosis increases patient's anxiety and social cost. Therefore, a timely CT can help both the physician and the patient by making an accurate diagnosis.

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사첨폐동맥판막과 폐동맥동맥류의 컴퓨터단층촬영 영상소견

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사첨폐동맥판막과 폐동맥동맥류가 함께 있는 것은 매우 드문 경우이다. 사첨폐동맥판막은 임상적으로 보통 큰 증상이 없어 대부분의 경우 사망 이후 발견된다. 그렇지만 최근 영상기술의 발달에 힘입어 우연히 사첨폐동맥판이 건강한 성인에서 발견되는 경우가 많아졌다. 이 증례보고에서는 임상적으로 무증상 성인에서 초음파에서 발견하지 못하고, 컴퓨터 단층촬영에서 발견된 사첨폐동맥판막과 폐동맥동맥류 증례를 보고하고 이전 선행연구들을 분석하고자 한다.

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