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- 1예 보고 -

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Congenital Quadricuspid Semilunar Valve

-A case report-

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A 17-year-old male patient was referred with symptoms of dyspnea. Multi-detector computerized tomography (MDCT) and echocardiography evaluation revealed quadricuspid aortic and pulmonary valves, an atrial septal defect (ASD), and pulmonary stenosis. We closed the ASD using a bovine patch and performed a commissurotomy of the pulmonary valve. Quadricuspid semilunar valves are very rare congenital abnormalities that are reported to occur nine times more frequently in the pulmonic valve than in the aortic valve. According to the Hurwitz and Roberts classification, the aortic valve was type A, and the pulmonic valve was type B. The aortic valve had normal function, but the pulmonic valve was stenotic and had abnormal function.

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Key words: 1. Valve disease
2. Pulmonary valve, stenosis
3. Heart valve, abnormalities

CASE REPORT

A 17-year-old male patient was referred with dyspnea which was aggrevated 3 months ago. A past history of mental retardation and polydactyly of the right foot were notable without the presence of chromosomal abnormality such as Down syndrome. The patient was diagnosed as having congenital heart disease, 14 years ago without proper treatment. The patient first came to our hospital two years ago with symptoms of dyspnea and was diagnosed with ASD, thickening of pulmonary valve, hypertrophy of the right ventricle, pulmonary hypertension and quadricuspid pulmonary and aortic valves through MDCT (Fig. 1).

On physical examination, systolic ejection murmur was auscultated on the left upper parasternal border, and there were no specific findings except right ventricle hypertrophy on electrocardiography. Mild hypoxemia was detected by arterial blood gas analysis; pH 7.423, pCO_2 44.0 mmHg, pO_2 70.5 mmHg, and sPO_2 94.4%. Right atrial and ventricular hypertrophy, atrial septal defect, and pulmonary stenosis were detected by echocardiography and maximal pressure gradient between right ventricle and main pulmonary artery estimated by Doppler echocardiography was 64 mmHg. Pulmonary valvular motion was restricted and the pressure gradient between right ventricle and main pulmonary artery was 60 mmHg by right atrial angiography. After diagnosing ASD and pulmo-

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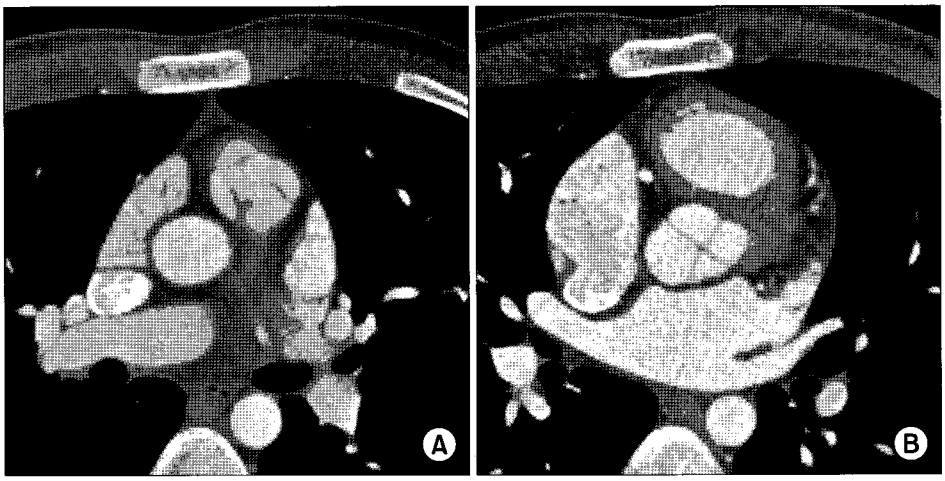

Fig. 1. Multi-detector computerized tomogram (MDCT) image showing both pulmonary (A) and aortic (B) quadricuspid valves.

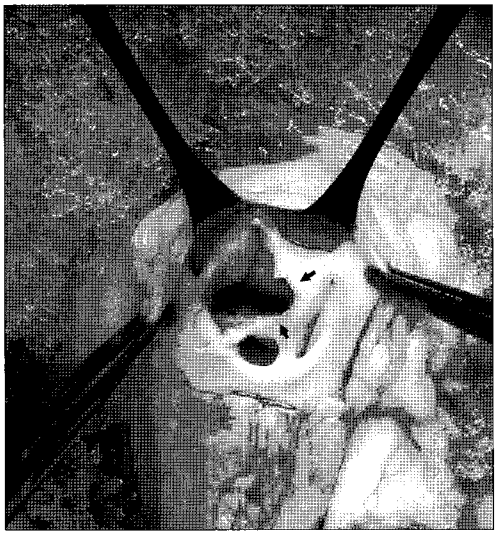

Fig. 2. Operative findings show quadricuspid pulmonary valve and 3-fused commissures. Commissurotomy of two site of the 3-fused commissures (arrow) was done.

nary stenosis, operation under routine cardiopulmonary bypass was performed.

After cardiopulmonary bypass, incisions to the pulmonary arteries were made. Quadricuspid pulmonary valve was confirmed under direct surgeon's visual field. Three parts of four commissures were fused. When performing commissurotomy on all three fused commissures, pulmonary regurgitation can occur. Therefore, commissurotomy was done on the left two commissures with 3 mm incision on each valve (Fig. 2). Afterwards right atriotomy and ASD closure to the verified 12 mm-size ASD was done using the bovine pericardial patch. The patient was discharged from the hospital on the seventh postoperative day without any complications. The aortic valve and pulmonary valve shows no abnormality on echocardiography at 11 months follow up after discharge.

DISCUSSION

Quadricuspid semilunar valves are very rare congenital malformations. To our knowledge, this is the first reported case of concomitant quadricuspid aortic and pulmonary valves. This abnormality appears to be due to abnormal process of truncus arteriosus dividing into aorta and pulmonary trunk during embryological development of the heart. Incidence of Quadricuspid pulmonary valve is 0.02~0.12% and that of quadricupid aortic valve is 0.008~0.043%, the former occurs about 9 times more often[1-5].

Developmental mechanism of quadricuspid valves had not been understood exactly, but Fernandez et al. explained the occurrance of quadricuspid valves by a hypothesis using embryo of a Syrian hamster. Normal aortic valve originates from three mesodermal primordial cell differentiation and one of these three mesodermal cells is detached by invagination of endothelial cell at valve development period, so this process results in the formation of quadricuspid valves[4].

Quadricuspid semilunar valves were classified into 7 types by Hurwitz and Roberts in 1973. Among the 121 cases, the majority of quadricuspid pulmonary valve was type B in 72 cases (60%)[2]. In this case, 4 cusps of aortic valve were same-sized, so called type A, pulmonary valve showed 3

same-sized commissures and the remained one cusp was small-sized, so called type B.

Most quadricuspid pulmonary valve functions normally, but conversely the quadricuspid aortic valve functions abnormally. The functional disorder of quadricuspid pulmonary valves shows regurgitation in most cases, but stenosis of the valves is rare[2,5].

Aortic valve and pulmonary valves were concomitant quadricuspid valves in our case and this is extremely rare. In contrast to previously reported properties of quadricuspid valves, the function of aortic valve was normal and pulmonary valves showed stenosis. Because the four cusps of the aortic valve were the same size, classified as type A, the aortic valve seems to function normally which is probably due to regular distribution of pressure[3,5].

In operation techniques to correct the abnormality, commisurotomy and valve replacement can be done. Commissurotomy of pulmonary valve was done after considering the patient's age and the possibility of re-operation. Post-operative echocardiography showed relieved pulmonary stenosis and no pulmonary regurgitation. Because the patient is still young and quadricuspid semilunar valves are unstable, periodical echocardiographical examination and regular follow-up to check other functional abnormalities in aortic and pulmonary valves are needed.

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=국문 초록=

호흡곤란을 주소로 내원한 17세 환자가 대동맥 및 폐동맥 사엽성 판막을 보이며 심방중격결손과 폐동맥 판막 협착증이 발견되어 소심막을 이용한 심방중격결손 봉합 및 폐동맥 판막 교련절개술을 시행하였다. 대동맥 판막과 폐동맥 판막이 모두 사엽성 판막을 보이는 경우는 매우 드문 선천성 기형으로 폐동맥 사엽성 판막이 대동맥 사엽성 판막보다 9배정도 많다. 본 증례는 Hurwitz and Roberts 분류법상 대동맥판막은 A형, 폐동맥판막은 B형이었으며 대동맥 판막기능은 정상적이었고 폐동맥 판막 협착증을 보였다.

중심 단어 : 1. 판막성 질환
2. 폐동맥판 협착증
3. 심장판막기형