

Tricuspid and pulmonary valve endocarditis associated with double-chambered right ventricle

Jin Cheol Myeong,¹ Jung Yeon Chin,¹ Jin Ho Choi,² Young Min Rah,¹ Jun Hyung Park¹

¹Department of Cardiology, Eulji University Hospital, Eulji University School of Medicine, Daejeon, Korea

²Department of Cardiovascular surgery, Eulji University Hospital, Eulji University School of Medicine, Daejeon, Korea

우심실 이분증과 연관된 삼첨판 및 폐동맥판 감염성 심내막염

명진철,¹ 진정연,¹ 최진호,² 나영민,¹ 박준형¹

¹을지대학교병원 심장내과학교실

²을지대학교병원 흉부외과학교실

We report a rare case of tricuspid valve and pulmonary valve endocarditis associated with a double-chambered right ventricle in an adult female with pulmonary artery aneurysm and septic pulmonary embolism by *Streptococcus mitis*. She was treated with aggressive antibiotic therapy followed by debridement of the infective lesion of tricuspid valve, pulmonary valve replacement using xenograft and resection of obstructing muscular bundles in right ventricle.

Key Words: Bacterial endocarditis, Congenital heart disease, Pulmonary valve, Right ventricle, Tricuspid valve

Double-chambered right ventricle (DCRV) is a rare grown-up congenital heart disease (CHD). It is characterized that RV is divided high pressure inlet and low pressure outlet by an anomalous muscle bundle.¹ In almost every case, DCRV is associated with other congenital

anomaly. In around 90 % of cases, perimembranous ventricular septal defect (pmVSD) is accompanied.² Echocardiographically, DCRV can be underdiagnosed because high velocity jet of RV outflow tract is misidentified for left to right shunt flow of pmVSD. The erroneous su-

Corresponding Author : Jin Cheol Myeong, Division of Cardiology, Eulji University Hospital, 95, Dunsan-Seoro, Seo-gu, Daejeon 302-120, Korea
TEL: +82-10-9976-9992 FAX: +82-42-611-3183 Email: spyman2000@naver.com

Received : Sep. 18, 2014

Revised : Oct. 8, 2014

Accepted : Oct. 15, 2014

ture of stenotic lesion of DCRV as a treatment of pmVSD could be resulted in RV failure and perioperative mortality.³ So, the accurate preoperative diagnosis of DCRV is essential. Pulmonary valve endocarditis is very rare accounting for less than 2% of admissions for endocarditis.⁴ The half of these cases occurred in patients with CHD. We here-in present a case of tricuspid and pulmonary valve endocarditis associated with DCRV which was successfully treated with antibiotic therapy and surgery.

Case

A 37-year-old woman referred for cardiac murmur from local clinic. She was suffered from cough with hemoptysis and dyspnea on exertion [The New York Heart Association (NYHA) Functional class II] for 1 month, not relieved by 2 weeks of antibiotic therapy (500 mg of Levofloxacin once daily). On physical examination, she was febrile (38°C), pale and had jugular vein engorgement. In auscultation, a harsh 4/6 grade systolic ejection murmur was heard along the 3rd left sternal border. Electrocardiogram showed right ventricular hypertrophy with strain pattern, right axis deviation, and right atrial enlargement (Fig. 1). Chest X-ray in the posteroanterior view revealed car-

diomegaly and multifocal patch consolidations in both lung fields. Laboratory findings revealed leukocytosis (white blood cell count; 17,389 x 10⁹/L with 81.6 % of neutrophils) and microcytic hypochromic anemia (hemoglobin 9.3 g/dL, mean corpuscular volume of 76.0 fl., serum iron 17 mcg/dL, transferrin saturation 6.2 %, serum ferritin 51.2 ng/mL) and elevated C-reactive protein 9.92 mm/h. Pulmonary angio computed tomography showed multiple pulmonary aneurysms with distal pulmonary infarction (Fig 2.). Mild cardiomegaly with right heart enlargement was also shown. Transthoracic echocardiogram (TTE) revealed hypertrophied RV septal and parietal muscular band and oscillating echogenic masses attached to tricuspid valve (6 x 6 mm to anterior leaflet and 11x8 mm to septal leaflet) and pulmonary valve (20 x 12 mm) (Fig. 3). Aneurysmal formation of perimembranous portion of ventricular septum was also observed but there is no definite shunt flow. Ceftriaxone (2 g/day i.v) with gentamicin (3 mg/kg/day i.v.) was started for treatment of infective endocarditis. In 8th day, *Streptococcus mitis*, a kind of oral (formerly viridans) streptococcus were grown in 3 bottles of initial intravenous blood sample. In 10th day, transesophageal echocardiogram (TEE) showed multiple elongated shaggy mass on hypertrophied muscular bands, suspicious vegetations additionally without size reduction of vegetations in tricuspid and pulmonary valve (Fig.4). In general,

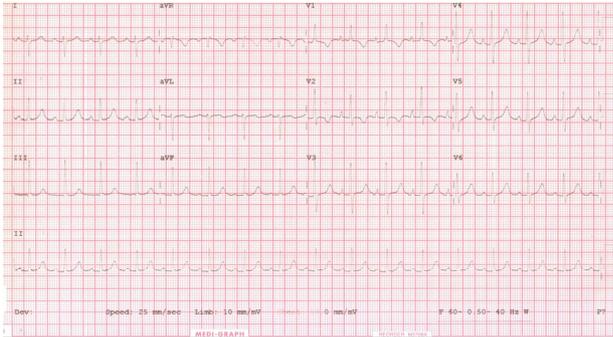


Figure 1. Initial electrocardiogram showing right ventricular hypertrophy with strain pattern, right axis deviation, and right atrial enlargement.

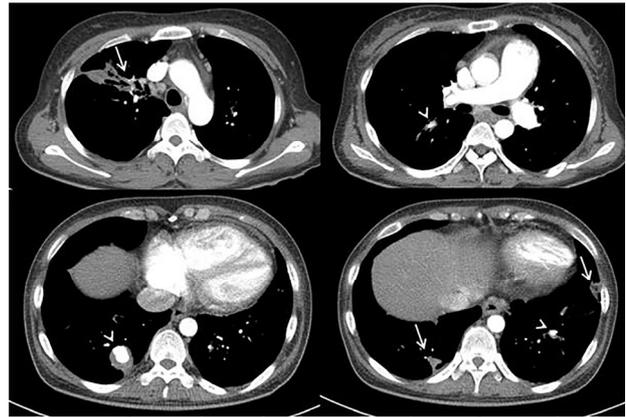


Figure 2. Pulmonary angio CT showing mild cardiomegaly with right heart enlargement and multiple distal pulmonary infarction (white arrow heads) with multiple pulmonary artery aneurysm (white arrows).

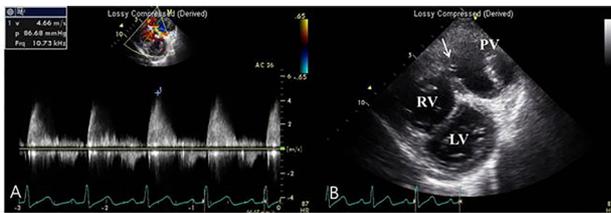


Figure 3. Transthoracic echocardiogram showing (A) continuous-wave Doppler tracing along the right ventricular outflow tract with peak gradient of 87 mmHg across the subinfundibulum (B) hypertrophied muscular bands dividing the right ventricle (white arrow) with D-shaped interventricular septum and large vegetation on pulmonary valve. LV: left ventricle, PV: pulmonary valve, RV: right ventricle.

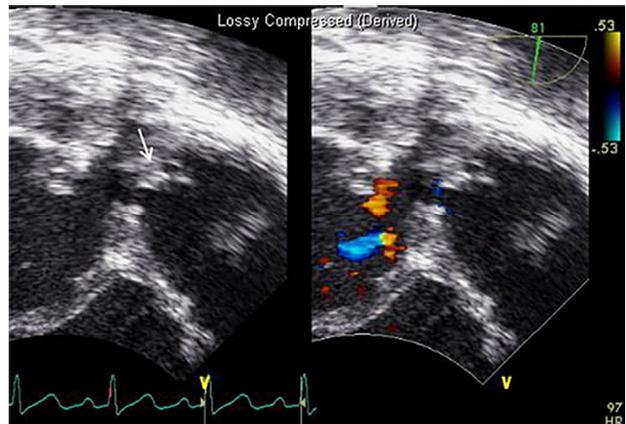


Figure 4. Transesophageal echocardiogram showing echogenic shaggy materials (white arrow) on hypertrophied muscular bands, suspicious vegetations.

intravenous antimicrobial therapy is recommended for IE. All symptomatic adult patients with DCRV undergo surgery to resect the obstructive muscle bands and to repair any associated lesions, because the obstruction is progressive among adults. So, the patient referred for cardiac surgery at 14th day. After right atriotomy, debridement of the infected area of tricuspid valve and hypertrophied muscular bands was performed. Excision of hypertrophic septal and parietal muscle bands causing infundibular stenosis was also done. The pulmonary artery was opened longitudinally. Pulmonary valve and vegetation was excised and replaced by xenograft (Edwards Prima™ Plus stentless Porcine Bioprosthesis). After cardiac surgery, her ECG showed newly developed complete right bundle branch block. Postoperative TTE showed no visible hypertrophied muscular band and no flow acceleration in the RV. For 4 weeks of intravenous antibiotics, she was discharged from the hospital.

Discussion

Double chambered right ventricle is a rare in adults in which is seen in only 0.5-2% of all cases of CHD.¹ It is characterized by an apical trabecular component of the RV, dividing the RV into a high pressure inlet chamber and a

low pressure outlet chamber. DCRV is associated with other cardiac anomalies in approximately 80-90% of patients, most frequently, pmVSD, or, less often, with membranous subaortic obstruction, valvular pulmonary stenosis, double-outlet RV, and atrial septal defect.² VSD incidence diminishes in adulthood. The echocardiographic diagnosis can be challenging, and is often misdiagnosed as obstructed ventricular septal defect, due to the high turbulent jets in the outflow tract of the RV.³ Also, in auscultations, both VSD and DCRV may have a systolic thrill and a harsh murmur easily audible along left sternal border.⁵ In reported cases, the stenotic area within the RV was erroneously sutured, since the lesion had been diagnosed simply as a VSD.⁶ Sometimes, patients with DORV had re-operation for DORV after surgical closure of VSD because of still remained turbulent flow in outflow tract of the RV. The treatment of choice is careful surgical resection of the obstructing muscular bundles. The time for intervention usually depends on the associated lesions. In the absence of a significant coexisting defect, observation may be appropriate as long as the intracavitary systolic gradient is not greater than 40 mm Hg and the obstruction is not progressive.¹ After surgery, nearly half developed complete right bundle branch block, but significant ventricular tachycardia was not detected in most cases. ³ Surgical treatment carries a very low risk and

provides excellent long term results.^{3,7} Right-sided infective endocarditis (IE) accounts for 5-10% of cases of IE.⁴ It is most frequently observed in intravenous drug abusers. IE in CHD is rare and more frequently affects the right heart. In this case, pulmonary valve was most likely initiated due to the impact of the accelerated jet originating from the muscular stenosis of RV. This was induced injuring the endocardial surface of the PV and TV, with subsequent thrombus formation over denuded valvular leaflet followed by infection.

In this case, TTE and/or TEE were valuable diagnostic tools in the treatment of DCRV with IE. Moreover, TEE showed that small vegetations attached to numerous myocardial fibers could be detected.

REFERENCES

1. Hoffman P, Wójcik AW, Rózański J, Siudalska H, Jakubowska E, Włodarska EK, et al. The role of echocardiography in diagnosing double chambered right ventricle in adults. *Heart* 2004;90:789-93.
2. López-Pardo F, Aguilera A, Villa M, Granado C, Campos A, Cisneros JM. Double-chambered right ventricle associated with mural and pulmonic valve endocarditis: description of a clinical case and review of the literature. *Echocardiography* 2004;21:171-3.
3. Telagh R, Alexi-Meskishvili V, Hetzer R, Lange PE, Berger F, Abdul-Khaliq H. Initial clinical manifestations and mid- and long-term results after surgical repair of double-chambered right ventricle in children and adults. *Cardiol Young*. 2008;18:268-74.
4. Habib G, Hoen B, Tornos P, Thuny F, Prendergast B, Vilacosta I, et al. ESC Committee for Practice Guidelines.: Guidelines on the prevention, diagnosis, and treatment of infective endocarditis (new version 2009): the Task Force on the Prevention, Diagnosis, and Treatment of Infective Endocarditis of the European Society of Cardiology (ESC). Endorsed by the European Society of Clinical Microbiology and Infectious Diseases (ESCMID) and the International Society of Chemotherapy (ISC) for Infection and Cancer. *Eur. Heart J* 2009;30:2369-413.
5. Choi YJ, Park SW. Characteristics of double-chambered right ventricle in adult patients. *Korean J Intern Med*. 2010;25:147-53.
6. Simpson Jr WF, Sade RM, Crawford FA, Taylor AB, Fyfe DA. Double chambered right ventricle. *Ann Thorac Surg* 1987;44:7-10.
7. Chaurasia AS, Nawale JM, Yemul MA. Double-Chambered Right Ventricle with Pulmonary Valve Endocarditis. *Echocardiography* 2013;30:167-70.