Diverticulum of the Right Ventricle Associated with Pulmonary Stenosis, Rhabdomyoma and Wolff-Parkinson-White Syndrome

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A true diverticulum of either ventricle is a very rare congenital cardiac anomaly, mostly seen as part of a syndrome of midline thoracoabdominal defect with other cardiac anomalies (so-called Cantrell’s syndrome), but rarely as an isolated malformation [1]. Most diverticula arise from the apex of the left ventricle. Right ventricular diverticulum is a rare condition that may originate in the apical or infundibular regions [2, 3]. We report a case of infundibular diverticulum of the right ventricle combined with pulmonary stenosis, right atrial rhabdomyoma, and Wolff-Parkinson-White (WPW) syndrome in a 9-month-old boy.

Abstract: Congenital cardiac diverticulum is a rare anomaly that may present as an isolated lesion or in association with other malformations. Diverticulum of the left ventricle is more common than that of the right ventricle. We report a case of cardiac diverticulum over the right ventricular outflow tract and associated pulmonary stenosis, right atrial rhabdomyoma, and Wolff-Parkinson-White syndrome in a 9-month-old boy. The delta wave disappeared after removal of the atrial rhabdomyoma.

Case Report

A 9-month-old boy was referred to our hospital for evaluation of a heart murmur. Physical examination revealed a well-developed infant with no stigmata of tuberous sclerosis. A grade 3/6 systolic ejection murmur was audible over the left upper sternal border. Echocardiogram disclosed mild valvular pulmonary stenosis with a pressure gradient of 37 mmHg between the right ventricle and main pulmonary artery, and a tumor mass located at the right ventricular septum (Fig. 1A). Twelve-lead surface electrocardiograms (ECGs) showed WPW syndrome of the right accessory pathway (Fig. 2A). During 10 months of follow-up, the child suffered from crying cyanosis and exertional dyspnea, which gradually progressed. Echocardiographic examination revealed pulmonary stenosis, the associated secondary infundibular stenosis became more severe and the tumor mass grew slightly. Cardiac catheterization revealed suprasystemic pressure in the right ventricle (106/8 mmHg). The right ventriculogram showed a dome-shaped pulmonary valve with marked hypertrophy of infundibular muscle and a diverticulum opacified at the right ventricular outlet tract (Fig. 1B).

Surgical excision of the right atrial mass, obliteration of the diverticulum, and right ventricular outlet tract reconstruction with the equinopericardium were performed. Histologic examination of the tumor revealed a rhabdomyoma. The WPW pattern on the ECG disappeared immediately after surgery (Fig. 2B). The patient remained well during 3 years of postoperative follow-up.

Discussion

Cardiac diverticulum should be differentiated from cardiac aneurysm. A cardiac diverticulum is characterized by an outpouched cardiac chamber, which consists of normal myocardium and contracts synchronously with the normal ventricles, as seen in this case. On the other hand, an aneurysm is a fibrous saccular lesion with paradoxical movement.
of the ventricle [3–5]. The embryologic events responsible for
the formation of ventricular diverticula are still unknown.
Abnormal development of the septum transversum and de-
flective attachment of the heart tube to the yolk sac have been
suggested to explain apical ventricular diverticulum [1].

The clinical presentations of cardiac diverticulum are
influenced by the presence of coexisting malformations. Most
patients with isolated diverticulum are asymptomatic, and
diagnosis is usually incidental, but the diverticulum can result
in embolism, arrhythmias, cardiac failure and rupture.
Therefore, surgical resection of the diverticulum was indi-
cated in this case, and would also be indicated for symptom-
atic patients or patients with other associated cardiac anomalies
[2–7].

A diverticulum originating from the right ventricle is much
more rare than one from the left ventricle. Right ventricle
diverticulum may arise from the apical or anterorsuperior
(infundibular) regions. The anterorsuperior form is usually
combined with congenital heart defects such as ventricular
septal defect, tetralogy of Fallot, double outlet of the right
ventricle, and pulmonary stenosis [2, 3]. The combined
malformations of right ventricular diverticulum, pulmonary
stenosis, rhabdomyoma and WPW syndrome have not been
reported previously.

Rhabdomyoma, a cardiac hamartoma, has occasionally
been reported in association with WPW syndrome, espe-
cially in patients with tuberous sclerosis [8]. As predicted
from the ECG algorithm, the accessory pathway in our patient
was located around the right posteroskeletal region, which was
also the location of the rhabdomyoma [9]. The delta wave
disappeared immediately after surgery, which supported this
correlation in our case.

![A) Echocardiogram (apical four-chamber view) showing a tumor mass at the
right side of the lower atrial septum (white arrowheads). B) Right ventriculo-
gram showing a diverticulum (black arrowhead) and val-
vular and infundibular pul-
monary stenosis. LA = left
atrium; LV = left ventricle;
RA = right atrium; RV =
right ventricle.

![Twelve-lead surface electrocardiograms showing the Wolff-Parkinson-White syndrome pattern of the right accessory pathway (A) and
Delta waves that disappeared immediately after surgery, with occasional premature atrial beats (B).]
References