Ruptured Anerysm of the Sinus of Valsalva Coexisting With a Ventricular Septal Defect and Single Coronary Artery — A Rare Combination —

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A 26-year-old man had been diagnosed with a cardiac murmur from birth. In 1998, he was admitted to hospital because of slight fatigue. A grade 5/6 continuous murmur was audible near the right sternal border at the second intercostal space. Doppler echocardiography detected an abnormal flow that suggested that an aneurysm of the right coronary sinus of Valsalva had ruptured into the right ventricle. Blood tests showed a 19% step-up in oxygen saturation value between the right atrium and right ventricle, indicating a ventricular septal defect with left to right shunt. Coronary angiography revealed a single coronary artery. Surgical repair was carried out and the patient made an uneventful recovery. This rare combination of a ruptured aneurysm of the sinus of Valsalva coexisting with a ventricular septal defect and a single coronary artery has not been reported previously. (Circ J 2003; 67: 470–472)

Key Words: Ruptured sinus of Valsalva aneurysm; Single coronary artery; Ventricular septal defect

ruptured sinus of Valsalva aneurysm (RSVA) is frequently associated with other congenital defects, particularly ventricular septal defects (VSDs). In these patients, the RSVA originates from the right coronary sinus and ruptures into the right ventricle.

Case Report

In August 1998, a 26-year-old man was admitted to hospital because of slight fatigue. He had a history of cardiac murmur from birth and in 1982 a ventricular septal defect (VSD) was diagnosed from cardiac catheterization and angiography. He did not have a history of smoking or use of alcohol or illicit drugs.

On admission, the patient was 180cm tall and weighed 65kg. He appeared well on physical examination. The pulse was 72beats/min and regular, and blood pressure was 132/50mmHg. The lungs were clear. A grade 5/6 continuous murmur was audible near the right sternal border at the second intercostal space. There was no peripheral edema.

The urine sample was normal, as were the results of hematology and blood chemistry. An electrocardiogram showed a normal rhythm at a rate of 70beats/min, with left ventricular hypertrophy. A chest radiograph (Fig 1) showed a normal cardiac silhouette, with a cardiothoracic ratio of 51%. The lungs were confirmed to be clear.

Transthoracic echocardiography showed a mildly dilated left ventricle with normal contractility. Doppler echocardiography detected an abnormal flow that suggested that a right coronary sinus of Valsalva aneurysm had ruptured into the right ventricular inflow tract (Fig 2).

Right cardiac catheterization revealed that the pulmonary capillary wedge pressure was 13mmHg, the pulmonary artery pressure was 35/10(20)mmHg, the right ventricular pressure was 41/18mmHg, and the right atrial pressure was 7mmHg. A blood sample showed a 19% step-up in oxygen saturation between the right atrium and right ventricle.

On the second day’s admission, supravalvular aortography showed the right coronary sinus of Valsalva aneurysm that had ruptured into the right ventricle (Fig 3). Coronary angiography revealed a single coronary artery originating from the left coronary sinus. The right coronary artery branched from the left anterior descending artery, and ran dorsally to the pulmonary artery. The coronary artery did not have significant stenosis (Fig 4).

Surgical repair was carried out in September 1998 using cardiopulmonary bypass. The thin-walled aneurysmal portion (6-7mm) was resected, and communication between the aorta and the right ventricle was closed by a pericardial patch.
patch. The VSD, which was type II (perimembranous defect), was also closed with a patch. The patient made an uneventful recovery.

Discussion

RSVA coexists with VSD in 30-60% of cases; namely, aortic insufficiency, pulmonary stenosis, aortic stenosis, coarctation of the aorta, patent ductus arteriosus, tricuspid insufficiency and left superior vena cava.1–4 The principal VSD associated with RSVA is the supracristal type (type I),5 but in the present case, the VSD was a perimembranous defect (type II).

In 1989, Chamsi-Pasha et al first described a case of RSVA that coexisted with a single coronary artery6 and in that same year, Goto et al reported another case.7 In 1991, Dazai et al described a case of an infective aneurysm of Valsalva complicated by a left single coronary artery8 and the year before, Cabrera et al had reported a case of a newborn infant with an aneurysm of the atrial appendage, ascending aorta and sinus of Valsalva associated to VSD, fibromuscular subaortic stenosis and a single coronary artery.9

A single coronary artery is encountered as an isolated finding in 0.024% of the population and is frequently associated with other congenital cardiac malformations.10 The combination of VSD and a single coronary artery has been reported in only 2 cases.11,12 Having a single coronary artery may predispose the subject to severe critical complications, such as acute myocardial infarction,13 but this was not observed in the present case.

To our knowledge, the combination of RSVA, VSD and single coronary artery has not been previously reported in the English literature. Aneurysms of the sinus of Valsalva are reported to rupture suddenly in 40% of patients; who then present with palpitations, fatigue, chest pain, dyspnea and cardiac insufficiency.1,2 The symptoms usually depend on the size of the aneurysm and the chamber into which it ruptures: for example, a RSVA draining into the venous chambers causes a sudden elevation in venous pressure and flow, which results in a decrease in aortic diastolic pressure.14 Although the RSVA occurred suddenly in the pres-
ent case, the symptoms were almost silent except for the slight fatigue. The reason for this is unclear and we speculate that it might relate to the small size of the RSVA. Another interesting finding was the fall in aortic diastolic pressure, which may have been the result of the RSVA draining into the right ventricle.

The long-term results are excellent after surgical treatment and the risk for recurrent fistula or VSD is low.2,14

References