A Huge Congenital Sinus of Valsalva Aneurysm Causing Cerebral Embolism and Hypoplastic Tricuspid Valve

Jong Jun Lee, MD¹, Wook-Jin Chung, MD¹, Sang-Jin Lee, MD¹, Sang Min Park, MD¹, Seok Jae Zeon, MD¹, Geum-Ha Kim, MD¹, Young Chan Jo, MD¹, Mi-Seung Shin, MD¹, Chul Hyun Park, MD² and Eak Kyun Shin, MD¹

¹Cardiology Division, Department of Internal Medicine, ²Department of Cardiovascular Surgery, Gachon University Gil Medical Center, Incheon, Korea

Congenital sinus of Valsalva (SOV) aneurysm is a rare cardiac abnormality. Rarely the aneurysm enlarges without rupture, cause symptoms of the mass effect by compressing the adjacent structures, obstruction of the right ventricular outflow with tricuspid regurgitation, infectious endocarditis, thrombus formation and myocardial ischemia/infarction. And SOV aneurysm can also be a source of embolism. We observed a patient with cerebral infarction in whom a huge SOV aneurysm, was diagnosed as the presumed source of cerebral embolism and the cause of hypoplastic tricuspid valve.

KEY WORDS : Sinus of Valsalva · Aneurysm · Cerebral infarction · Tricuspid valve.
Chest X-ray showed a large rounded mass emerging from the mediastinum, towards the right side (Fig. 1B). Computerized tomography (CT) scan of the brain showed a low density area in the left parietal lobe suggesting left middle cerebral artery territory ischemic stroke (Fig. 1C). Biochemical and hematological examination were performed. And the values are as follows: hemoglobin 15 g/dL, hematocrit 43.4%, WBC 6,220/mm³, segment neutrophil 58.7%, platelet 142,000/mm³, PT 13.9 sec, AST 83 U/L, ALT 78 U/L, and gamma-GT 64 U/L. Other variables including the cardiac enzyme, the lipid profile and VDRL were within normal limits. Transthoracic echocardiograms demonstrated a huge cystic structure (6.27 cm × 4.61 cm) at the non-coronary sinus, which compressed both atria (Fig. 2). On Doppler study, other structural disorders, including aortic regurgitation and ventricular septal defect (VSD) were not seen. Additionally tricuspid valve function could not be evaluated because of huge mass. Although the urgent operation was recommended, it was postponed considering acute phase of cerebral infarction. After a month, operation was performed via median sternotomy and serial intraoperative transesophageal echocardiographies were obtained during operation. Transesophageal echocardiogram (TEE) before cardioplegia confirmed huge SOV aneurysm (6.5 cm × 4.6 cm × 7.0 cm) and also showed severe swirling within the aneurysmal sac (Fig. 3). But, there was no evidence of current or previous endocarditis. After complete removal of aneurysmal sac, and patch repair of aneurysm opening by autologous pericardium was done. Although there was no deformity of tricuspid annulus, hypoplasia of anterior and septal leaflets of tricuspid...

Fig. 5. Intraoperative transesophageal echocardiography after heart reopening and tricuspid valve repair. A: Midesophageal short-axis view showed decreased size of right atrium and improved interatrial septum bulging. B: Color Doppler recording in the midesophageal four chamber view showed still remained grade II/IV of tricuspid regurgitation. C: Color Doppler recording in bicaval view also showed grade II/IV of tricuspid regurgitation. RV, right ventricle; RA, right atrium; LV, left ventricle; LA, left atrium; Ao, aorta.

value was seen. Intraoperative TEE showed grade III/IV of tricuspid regurgitation (TR) and bulging of interatrial septum (IAS) to the left side, which meant markedly elevated right atrial pressure. So, we decided to perform tricuspid valve repair (Fig. 4). After tricuspid valve repair, intraoperative TEE showed no more bulging of IAS but newly developed hematoma (2.4 cm × 1.62 cm) within IAS. TR was improved but still remained as grade II/IV (Fig. 5). After operation, her vital sign was unstable and right ventricular assist device (RVAD) was applied. However right ventricle failure ensued and eventually patient was expired at the day after operation.

**DISCUSSION**

John Thurnam first described sinus of Valsalva aneurysm in 1840 and Hope further described it in 1939. In Korea, Lee et al. also described 6 cases of SOV aneurysm rupture and operation. The prevalence of unruptured SOV aneurysms is difficult to assess, because they rarely cause symptoms and may even be missed at necropsy.

SOV aneurysm is more common in oriental people, about fivefold than in western people and the male to female ratio is approximately 2:1. Because of the relative infrequency of SOV, investigators have not accurately determined the natural history of the lesion. SOV aneurysm is considered as either congenital or acquired form of origin. Acquired form was secondary to syphilis, bacterial endocarditis, atherosclerosis, aortic dissection, Marfan’s syndrome, Behcet’s disease or trauma. It is most commonly originated from the right aortic sinus (90%) and non-coronary sinuses (8%), and only 2% arises from the left aortic sinus. The most common coexisting congenital heart diseases are ventricular septal defect, usually subaortic (25-55%), regurgitation of the aortic valve and rarely associated with pulmonary stenosis, patent ductus arteriosus, atrial septal defect, subaortic stenosis and tetralogy of Fallot. In this case, evidence of arteriosclerosis, a history of hypertension, signs of Marfan’s or Ehlers-Danlos syndrome or laboratory evidence of syphilitic infection were all absent. So we thought this case maybe a congenital from.

Unruptured SOV aneurysm can cause obstruction of the right ventricular outflow with tricuspid regurgitation, infectious endocarditis, thrombus formation with subsequent embolic events and myocardial ischemia/infarction due to compression of the stem or main branches of left coronary artery by the body of the aneurysm of the left sinus or obstruction of the ostium of the right coronary artery by thrombus. In this case, we thought that cerebral infarction and hypoplastic tricuspid valve caused by congenital aneurysm of SOV.

To the date, this case is the first case of a huge SOV aneurysm associated with cerebral infarction and fatal hypoplastic tricuspid valve abnormality. As a matter of course, an unruptured SOV aneurysm should be included in the list of sources of embolism. And also as huge congenital SOV aneurysm can cause tricuspid valve hypoplasia which may result in fatal condition, it should be considered for surgical plan of SOV aneurysm.

**REFERENCES**