中文題目：病例報告：主動脈竇瘤破裂至右心房所致難治性肺水腫

英文題目：Case Report: Refractory Pulmonary Edema Resulted from Ruptured Sinus of Valsalva Aneurysm to Right Atrium

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Background:

A ruptured sinus of Valsalva aneurysm is a rare cardiac abnormality and is associated with a severe left-to-right shunt if communicating with the right-sided heart chambers. In one-third of patients, left-to-right shunting immediately following the rupture of an aneurysm into the right side of the heart produces acute dyspnea and chest pain. However, half of patients note gradually worsening dyspnea, fatigue, chest pain, and peripheral edema over several months or even years following rupture, and the remainder of patients are still asymptomatic at the time of diagnosis. If left untreated, sinus of Valsalva aneurysm may cause severe heart failure, and even mortality. We present a case of patient with refractory pulmonary edema due to a ruptured sinus of Valsalva aneurysm.

Case Presentation

This 75-year-old man with previous history of chronic obstructive pulmonary disease (COPD) was admitted via emergent room (ER) because of aggravated dyspnea, orthopnea, leg edema for 1-2 weeks. He was diagnosed as COPD, severe and treated at local hospital for months. But, symptoms still progressed. Therefore, he was referred to our hospital.

At ER, the temperature was 36.8°C, the blood pressure 145/63 mm Hg, the pulse 88 beats per minute, the respiratory rate 22 breaths per minute, and the oxygen saturation 99% while he was breathing ambient air. Physical examination revealed regular heart beats, grade 3/6 systolic murmur over left lower sternal border (LLSB), and apex; grade 2/6 diastolic murmur over bilateral 2nd intercostal space and LLSB; bilateral rales over all lung field; bilateral pitting edema 3+. Cardiomegaly with pulmonary edema, mild bilateral pleural effusion was noted by chest radiography. Electrogram showed complete right bundle branch block. Laboratory studies showed normal CBC, normal troponin I and albumin, mild elevating creatinine, NT-proBNP. Echocardiography revealed adequate left ventricular ejection fraction; moderate to severe mitral regurgitation (MR); severe tricuspid regurgitation (estimated pulmonary artery systolic pressure: 58mmHg); pulmonary regurgitation; a jet from near aortic sinus to right atrium was suspicious. To clearly differentiate was difficult. Heart failure with pulmonary edema was diagnosed. We tried diuretics and spironolactone for fluid overload, valsartan for severe MR, theophylline for COPD. Orthopnea did not improve until high dose intravenous (IV) Furosemide (20mg q12H →40mg, q6H). He received cardiac catheterization for severe MR survey and possible intracardiac shunt survey.

Cardiac catheterization revealed patent coronary artery. A jet from sinus of Valsalva to right atrium was noted by aortogram. Shunt at atrial level (SVC/IVC to RA) was detected by oximetry run. Qp/Qs:1.74. Ruptured sinus of Valsalva aneurysm with left to right shunt was diagnosed.

Transesophageal echocardiogram (TEE) also confirmed this diagnosis. Early surgical intervention or transcatheter closure was the treatment of choice.

After discharged, he received oral diuretics. Body weight gained, and pulmonary edema recurred, so that he was hospitalization twice during the following 1 year. The symptoms got improving after IV diuretics. Then, elective transcatheter closure for ruptured sinus of Valsalva aneurysm was performed. The following echocardiography showed no residual shunt 2 months later. Besides, heart failure with pulmonary edema had not recurred under low dose diuretics since then.

Discussion and Conclusion：

Sinus of Valsalva aneurysm (SVA) is uncommon and is encountered in 0.14–0.96% of open-heart surgery cases. The incidence of SVA is reported to range between 0.1 and 3.5% of all congenital heart defects. The incidence of ruptured SVA is five times higher in Asian countries (0.46-3.5% in Eastern and 0.14-0.23% in Western areas). Of the group, 65-80% are males (male/female ratio 4:1). Diagnosis can occur at any age (average: 39 years, range: 2-74 years). Most ruptures develop between 20 and 40 years of age. This patient was diagnosed in older age.

The aneurysms usually arise from the right coronary sinus (65-85%), less frequently from the noncoronary sinus (10-20%) and rarely from the left coronary sinus (<5%).

Although an unruptured aneurysm of the sinus of Valsalva is usually asymptomatic, it may become symptomatic when it ruptures into one of the cardiac chambers. Symptoms can range from angina to acute pulmonary edema and cardiac collapse. In this patient, he noted gradually worsening dyspnea, and peripheral edema over several months.

If left untreated, SVA may cause severe heart failure, and even mortality. Surgical repair or percutaneous closure is therefore indicated.

Sinus of Valsalva aneurysm most commonly ruptures into the RA or RV which are low-pressure chambers producing a continuous murmur. Murmur is classically louder in diastole and this feature can be used to differentiate from the murmur of PDA or arteriovenous (AV) fistula. It is generally located along the left parasternal border with the site of loudest murmur and thrill suggesting the possible location of rupture. Systolic suppression of murmur may occur because of mechanical narrowing of fistulous tract during systole, and the murmur may be only diastolic in case of rupture into the left ventricle (LV).

In summary, ruptured sinus of Valsalva aneurysm might be a cause of pulmonary edema. Percutaneous closure of a ruptured sinus of Valsalva aneurysm is feasible and improves pulmonary edema in this kind of patient.