A 77-year-old man was admitted for congestive heart failure. He had started to have palpitations after eating supper 2 days before the admission and had experienced dyspnea on exertion since then. He visited his family physician, who referred him to the cardiology clinic of our hospital. The patient was under the family physician’s care for diabetes mellitus, hypertension, and paroxysmal atrial fibrillation. He had never been diagnosed as having any organic heart disease. He was an ex-smoker and drank alcohol on social occasions. The patient was on β-blockers and Coumadin.

His cardiovascular physical examination was significant for a 3/6 systolic murmur and a rapid, irregular heart beat. His ECG showed atrial fibrillation with a ventricular rate of 150 bpm. His portable chest x-ray (Figure 1A) showed bilateral pleural effusion and pulmonary edema. An emergency trans-thoracic echocardiogram showed normal left and right ventricular function. A large mass in the left atrium was seen that was causing functional mitral stenosis. The patient was admitted to the hospital, given digoxin, and started on intravenous diuresis. His ECG on normal sinus rhythm (Figure 1B), which was recorded 2 weeks after the admission, showed P mitrale, suggesting significant overload of the left atrium.

A complete transthoracic echocardiogram after conversion to normal sinus rhythm revealed a large aneurysm in the left coronary sinus of Valsalva measuring 58×48 mm that was protruding into the left atrium. There was a thrombus inside the aneurysm. The aneurysm was prolapsing into the mitral valve, thus resulting in obstruction of the left ventricular inflow. The mitral valve orifice area measured by the pressure–half-time method was 0.8 cm². No mitral regurgitation was observed at this time. The estimated right ventricular systolic pressure was 47 mm Hg. Significant tricuspid regurgitation was present. Aortic regurgitation was trivial. Transesophageal echocardiography showed the same pathophysiological findings (Movie I in the online-only Data Supplement).

The patient was further evaluated by computed tomography of the ascending aorta and the heart (Figure 2). This imaging modality further confirmed the findings from the echocardiogram.
At first, the patient did not oppose the idea of undergoing surgery and was willing to have preoperative diagnostic tests. However, he changed his mind when the heart failure symptoms resolved after heart rhythm conversion and diuresis. He refused surgery and continued to refuse it for the next 3 years, despite recommendations by the medical staff and recurrent hospitalizations for congestive heart failure.

He finally hoped to undergo surgery for exacerbating symptoms of chronic heart failure but died of pneumonia during the waiting period. His family did not want an autopsy to be performed. The last transthoracic echocardiography data recorded 3 months before his death revealed an expanded aneurysm and severe mitral regurgitation, which had not been observed at his first admission (Figure 3 and Movie II in the online-only Data Supplement).

Aneurysms of the sinus of Valsalva are uncommon, with an incidence ranging from 0.1% to 3.5% of all congenital heart defects and with a reported prevalence of 0.09% in an autopsy series. Sinus of Valsalva aneurysms are found most often in the right coronary sinus, less often in the noncoronary sinus, and least often in the left coronary sinus.

Aneurysms of the sinus of Valsalva are usually diagnosed in the setting of clinical sequelae of an acute rupture into adjacent cardiac structures. Although most unruptured sinus of Valsalva aneurysms remain asymptomatic, multiple case reports describe unruptured sinus of Valsalva aneurysms causing significant anatomic and physiological derangement and related symptoms. They may present with conduction-system abnormalities, thromboembolism, and myocardial ischemia resulting from an obstruction of the coronary flow. Protruding sinus of Valsalva aneurysms can also cause an obstruction of the cardiac chambers and valve insufficiency by distorting cardiac valves. Multiple reports have described right ventricular outflow obstruction. Rare cases of left ventricular outflow obstruction caused by right sinus of Valsalva aneurysms have been described. Right ventricular inflow obstruction has also been reported. However, to the best of our knowledge, mitral obstruction by a Valsalva aneurysm has never been described in the English literature. This may be related to the relative infrequency of aneurysms in the left sinus of Valsalva.

The direct cause of death in this patient was pneumonia. However, the patient showed progressive cardiac cachexia during the 3 years that he refused surgery. Early surgery is therefore strongly recommended for this unusual condition.

Disclosures

None.

References

Figure 3. Transthoracic echocardiography recorded 3 years after the first admission. The patient died of pneumonia 3 months after this study. Long-axis (A) and short-axis (C) views showed the large aneurysm in the left sinus of Valsalva (60×48 mm) that protruded into the left atrium and obstructed the mitral valve. Color Doppler mapping of the long-axis view (B) revealed severe mitral valve regurgitation, which had not been observed at his first admission. Ao indicates aorta; LA, left atrium; LV, left ventricle; R, right coronary sinus; N, noncoronary sinus; and L, left coronary sinus. *Sinus of Valsalva aneurysm.
Unruptured Left Coronary Sinus of Valsalva Aneurysm Causing Mitral Valve Obstruction
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