Acute Aortic Regurgitation Caused by Spontaneous Aortic Valve Rupture

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We report a rare case that manifested as severe aortic regurgitation (AR) resulting from spontaneous aortic valve (AV) rupture caused by valvular myxomatous transformation in a middle-aged woman. Before operation, a ruptured hole on the left coronary cusp (LLC) was clearly visualized with transesophageal echocardiography (TEE). There were no clinical findings of vegetations or sinus Valsalva aneurysms. The patient was treated successfully with mechanical AV replacement. Prompt imaging evaluation is mandatory in this rare critical situation for correct diagnosis and appropriate management.

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A 51-year-old woman without other medical conditions (with the exception that she was a smoker) presented to a local hospital with progressive dyspnea on exertion of 1 week’s duration. She was transferred to our medical center after immediate intubation for acute respiratory distress. On admission, her blood pressure was 140/60 mm Hg, pulse rate was 90 beats per minute, temperature was 37.5°C, and respiration rate was 20 breaths per minute with support of a ventilator. The physical examination revealed a grade III/VI early diastolic murmur along the left sternal border with a loud pulmonary component of the second heart sound. The chest roentgenogram showed a bilateral ground-glass pattern. Transthoracic echocardiography (TTE) disclosed severe AR caused by increased left ventricular stroke volume and widened pulse pressure. The left ventricular end-diastolic pressure was elevated and equalized with the aortic end-diastolic pressure. The continuous wave Doppler showed markedly decreased pressure half-time. All these clues pointed to a more acute course of AR, although a preexisting mild AR with an acute progression of severity could not be completely ruled out regarding the borderline left ventricular end-diastolic dimension.

Comment

In such a case presenting as frank pulmonary edema and severe AR, the first diagnostic challenge for clinicians is to determine the course of the AR because there are different causes and treatment strategies. Our patient was totally symptom free before this event. She had a soft early diastolic murmur without typical signs of chronic severe AR caused by increased left ventricular stroke volume and widened pulse pressure. The left ventricular end-diastolic pressure was elevated and equalized with the aortic end-diastolic pressure. The continuous wave Doppler showed markedly decreased pressure half-time. All these clues pointed to a more acute course of AR, although a preexisting mild AR with an acute progression of severity could not be completely ruled out regarding the borderline left ventricular end-diastolic dimension.

In clinical situations of acute severe AR, aortic dissection, infective endocarditis, and sinus of Valsalva aneurysm rupture are on the list of differential diagnoses. Lacking the evidence supporting these diagnoses, AV rupture should be considered. Although one might be impressed by the prolapsed pattern of the
AV in our case when first viewing the echocardiographic images, searching for the underlying mechanisms causing the valvular prolapse is more critical. The transesophageal images in our case demonstrated the striking finding of a ruptured defect on the LCC, which indicated to us this rare diagnosis. Without a history of trauma and infectious processes, spontaneous AV rupture was favored to be the cause of the acute severe AR in our patient.

The pathologic link between spontaneous AV rupture and myxomatous transformation has scarcely been mentioned in the literature. We were able to find only 5 cases in a review of the literature [1–4]. In an era that lacked 2-dimensional echocardiography, the M-mode images served as the diagnostic tool [1, 2] to demonstrate the prolapsed AV as thick bands of echoes occupying the aortic root throughout diastole or as the abnormal echo-genicity in the left ventricular outflow tract; this simulated the M-mode findings in our case. In the present case, we hypothesized that the myxomatous degeneration of the AV led to the prolapse. The combination of the progress of the disease per se and the effect of the shear force of the blood flow created a weak point wherein the rupture occurred. Consequently, the AR jet dragged the AV down and resulted in a more severe prolapse, thus begetting a more severe AR.

In conclusion, in the absence of clinically identifiable secondary causes for acute AR, such as infective endocarditis, aortic dissections, sinus Valsalva aneurysm rupture, or trauma, an uncommon diagnosis of spontaneous AV rupture should be considered and recognized. Surgical intervention should not be delayed, and
intraoperative tissue pathologic examination is important to provide an insight into the mechanisms of this rare disease. Prompt imaging evaluation, especially TEE, is also crucial in such a critical situation for correct diagnosis and appropriate management.

References