Ruptured sinus of Valsalva aneurysm as the cause of death of an 18-year-old student

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A sinus of Valsalva aneurysm (SOVA) is very rare, with the general population prevalence estimated at 0.09% [1]. SOVA is usually asymptomatic before rupture, which typically occurs in early adulthood and may have various clinical manifestations [1–4].

We report a case of an 18-year-old woman who suffered from symptoms of predominantly right heart failure for 6 months and was hospitalized twice for ascites and pleural effusion (gynecology and internal ward). Eventually, she was urgently admitted to our cardiology ward with suspicion of SOVA based on an outpatient echocardiogram. On admission, she presented with NYHA class II and no symptoms of fluid retention. On physical examination, a continuous cardiac murmur was found, loudest at the left parasternal border (5/6). Echocardiography revealed a ruptured aneurysm of the noncoronary sinus of Valsalva with a shunt into the right atrium and enlargement of the right heart chambers (Figure 1A, 1C). The diagnosis was confirmed by cardiac CT (Figure 1B). On the fourth day, when the Heart Team consultation was planned to qualify for cardiac surgery, the patient's clinical condition suddenly deteriorated, and a sudden cardiac arrest occurred with a third-degree atrioventricular block observed on the cardiac monitor. After resuscitation and return of spontaneous circulation, the patient was secured with an electrode for temporary cardiac pacing and transferred to the Intensive Care Unit. The patient was qualified for urgent cardiac surgery provided that her condition would allow transport to another hospital. Despite intensive efforts, symptoms of cardiogenic shock and multi-organ failure increased. The patient suffered further cardiac arrests and despite prolonged resuscitation died.

An autopsy was performed. Macroscopically, the coronary sinus of the Valsalva aneurysm was confirmed (Figure 1D) with a transverse, linear full-thickness wall fracture (1 cm of length) visible at its apex (Figure 1B), communicating with the right atrium. Right heart cavity wall hypertrophy and dilatation, as well as transudative fluid in the pleural cavity, peritoneal cavity, pericardial sac, congestive lesions in the lungs, spleen, and nutmeg liver, were revealed. Microscopically, a transmural aneurysm without signs of wall delamination or fresh destruction advanced hemosiderosis and massive congestion of the liver was found. The cause of death was assumed to be cardiogenic shock in the course of right ventricular heart failure caused by leakage from a ruptured SOVA.

Surgery of a ruptured SOVA is performed with acceptably low mortality (1.9% to 3.6%) and good long-term outcome (survival rate is up to 90% at 15 years) [3]. The prognosis in a ruptured untreated SOVA is poor — death usually occurs within 1 year due to congestive heart failure [2]. Longer survival times have been described only in patients with slow perforation (mean survival period up to 3.9 years) [5]. Therefore, in cases of a ruptured SOVA, urgent surgery should be performed. However, the exact definition of that urgency has not been specified, mostly due to the literature being largely based on casuistry. Our case illustrates that ruptured SOVA is truly a life-threatening condition that requires immediate surgical intervention following minimal and necessary imaging.

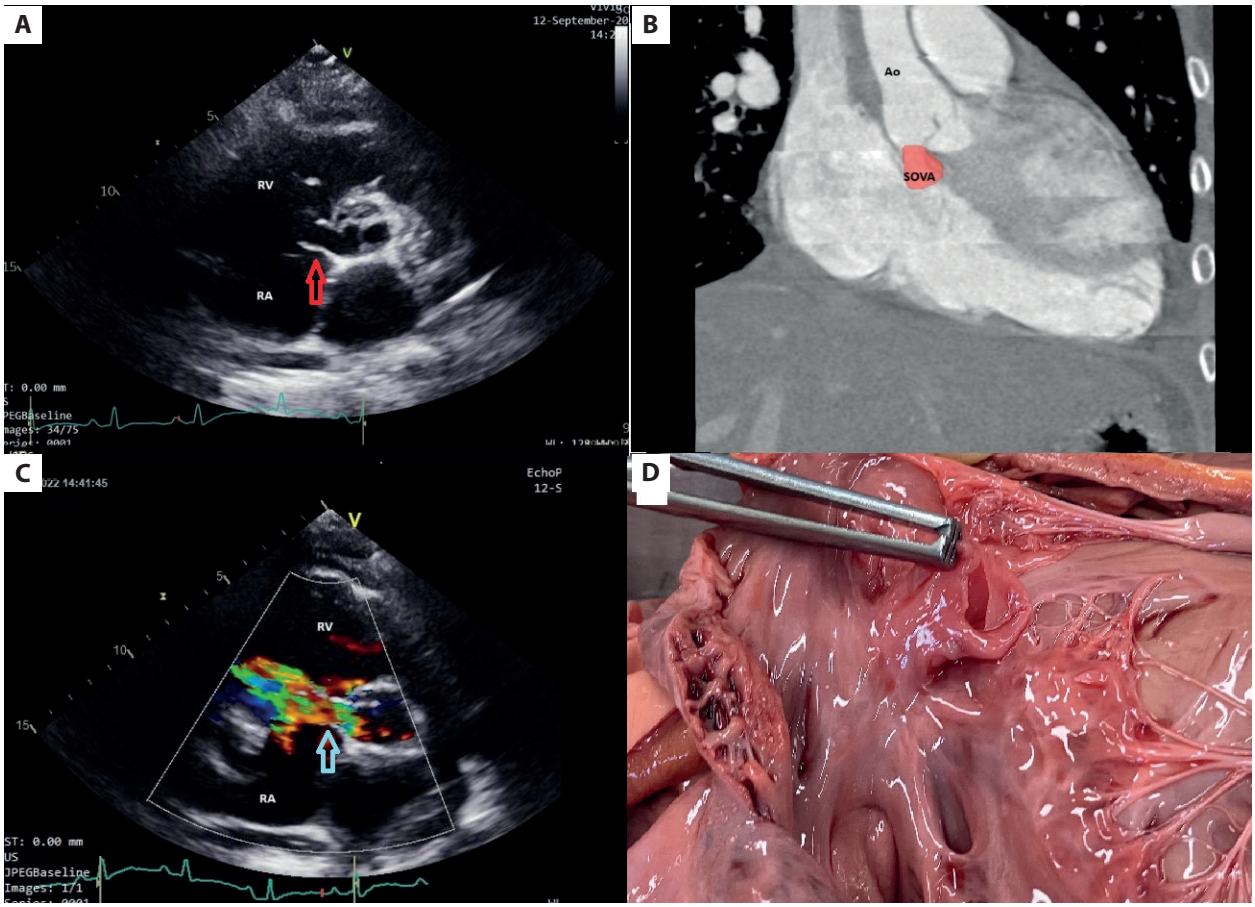

Figure 1. A. Transthoracic echocardiography (SAX) shows a ruptured aneurysm of the right coronary sinus of Valsalva (red arrow). **B.** Cardiac computed tomography shows a ruptured aneurysm of the right coronary sinus of Valsalva. **C.** SAX with color Doppler shows blood flow from the noncoronary sinus to the right atrium (blue arrow). **D.** Autopsy examination: Ruptured coronary sinus of Valsalva aneurysm

Article information

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