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Angina pectoris and sinus of Valsalva aneurysm: a rare manifestation of an uncommon disease

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Sinus of Valsalva aneurysm is an uncommon finding. The present paper describes a case report of a 57-year-old female patient who developed angina pectoris due to the left sinus of Valsalva aneurysm. Clinical presentation, localization, unruptured status and concomitant coarctation of the aorta make the case presented in this document extremely rare and interesting. Current knowledge about the disease is briefly discussed.

Keywords: angina pectoris, sinus of Valsalva aneurysm, congenital heart disease, aortic coarctation

INTRODUCTION

Angina pectoris is a common medical problem worldwide. The 2017 Heart Disease and Stroke Statistics update of the American Heart Association reported that 16.5 million people aged 20 and above in the United States have coronary heart disease (CHD) (1), therefore the prevalence of angina pectoris syndrome is high. Although the common understanding of pathophysiology of angina pectoris is closely related to atherosclerosis (2), there are other causes of myocardial ischemia (3) that need to be considered. An extremely rare cause of myocardial ischemia is sinus of Valsalva (SoV) aneurysm that can impair coronary circulation by compressing the epicardial coronary artery and disturbing the blood flow (4). The present case report describes an interesting clinical case of angina pectoris related to SoV aneurysm and addresses the importance of pre-invasive examination of suspected CHD patients. Contemporary issues of SoV aneurysm are briefly discussed.

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Case report

A 57-year-old female patient complaining of angina-type chest pain was consulted by a cardiologist in an outpatient setting. The patient was diagnosed with arterial hypertension 25 years ago and denied any other chronic diseases. As her rest ECG was normal, exercise ECG test was performed after CHD was suspected. The test was positive; ST depression of 2 mm in leads V1–V4 was provoked by physical activity. 2D echocardiography was performed by a local cardiologist and revealed no significant abnormalities. The patient

was referred to Klaipėda Jūrininkai Hospital interventional cardiology centre for an interventional coronary angiography procedure. The radial artery approach was chosen to reach the SoV. Since the attempts to find the opening of the left main coronary artery (LMCA) were unsuccessful, unselective contrast aortography was performed to identify the anatomy of the left sinus of Valsalva (LSoV). It was then observed that the LSoV was severely dilated. The right coronary artery was visualized in a typical position and no pathologic changes were noticed. Echocardiography was then repeated at our institution. The investigation revealed that the LSoV was severely dilated, therefore, LSoV aneurysm (40 × 37 mm) was suspected (Fig. 1). Left ventricular hypertrophy with excellent left ventricular systolic function (EF >65%) and impaired relaxation was also noted. Mild aortic and mitral valve insufficiencies were detected. Computed tomography (CT) angiography of aorta and ECG-gated cardiac CT angiography were performed and showed aortic coarctation just after the left subclavian artery with aortic narrowing up to 9 mm (Fig. 2) and LSoV aneurysm (45.5 × 36.9 mm) with LMCA involvement (Figs. 2, 3). The opening of the LMCA was rising from the aneurysm sac and the proximal portion of the vessel was lying on the body of the aneurysm (Figs. 2, 3). No atherosclerotic changes were detected in the coronary arteries. All laboratory markers including test for syphilis were negative. Valve-sparing open surgery was chosen. Aortic prosthesis was used and a tongue-shaped extension of the prosthesis tube was formed to cover

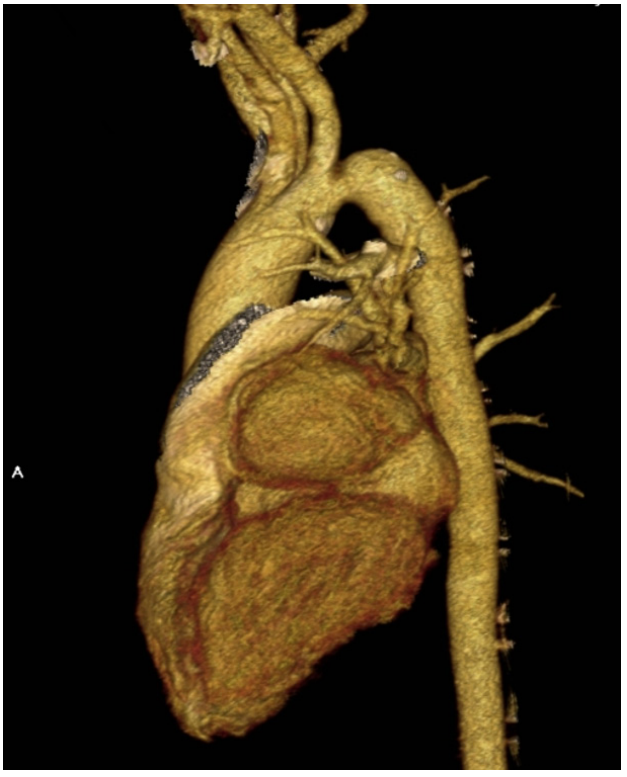

Fig. 2. Three-dimensional reconstruction of cardiac chambers and aorta: aortic coarctation up to 9 mm behind the left subclavian artery is visualised; the left sinus of Valsalva aneurysm and the proximal portion of the left main coronary artery is reconstructed

the resected defect. The LMCA was reimplanted into the newly-formed sinus. The patient was asymptomatic for coarctation, therefore no surgical intervention was performed to treat it. Histologic examination of the resected tissue represented true aneurysm with no specific findings. No surgery-related complications developed. Surgical

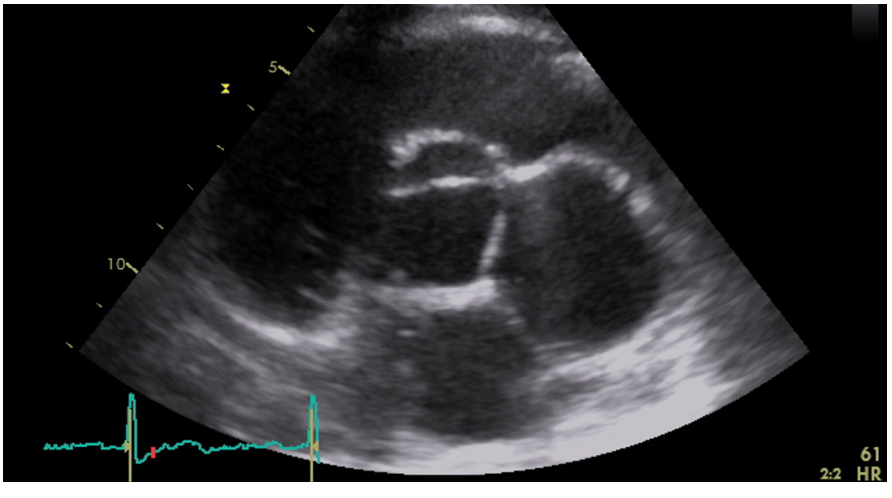

Fig. 1. Parasternal short axis (aortic) view: the left sinus of Valsalva aneurysm

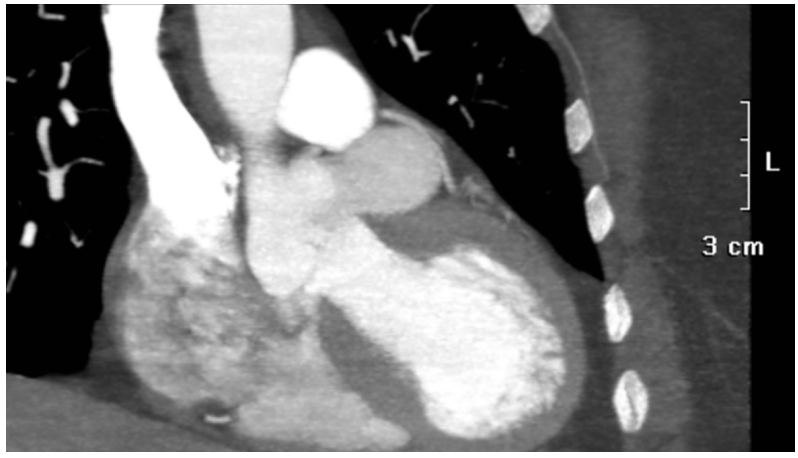

Fig. 3. ECG-gated cardiac CT angiography: the left sinus of Valsalva aneurysm gives rise to the left main coronary artery which then lies on the aneurysm body

treatment was effective in restoring normal coronary flow; the patient was free from angina-type chest pain and dyspnea.

DISCUSSION

Pathologic dilation of aortic sinuses can be a consequence of either SoV aneurysm or pseudoaneurysm (false aneurysm). Since both abnormalities are uncommon, true prevalence remains unknown. The autopsy data revealed that these pathologies are found in 0.09% of subjects (5). However, the rate of incidence is highly dependent on the population under analysis. It is noted that men are more often affected than women and Asian descent is a risk factor (6, 7). It has been shown that the right sinus of Valsalva (RSoV) is affected most often. It accounts for 70–90% of SoV aneurysm cases. The aneurysm of non-coronary SoV is diagnosed much less often (10–20% of cases). Aneurysms of the LSoV are exceedingly rare (5, 7). Yan et al. analysed 160 cases of SoV aneurysms and concluded that the prevalence of LSoV aneurysm is less than 1% (8). Moreover, it is also noted that unruptured SoV aneurysm is diagnosed only in 5% of cases (8). Clinical presentation, localization, the unruptured status, and concomitant coarctation of the aorta make the case presented in this paper extremely rare and interesting.

It is difficult to draw a precise picture of the causes because SoV aneurysms are rare and the majority of literature comes from individual case reports or surgical case series. SoV aneurysms are often attributed to congenital pathologies. Congenital diseases that can lead to SoV aneurysm are Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, and other rare connective tissue disorders (5, 6). On the other hand, different acquired pathologies can also weaken the aortic wall and lead to dilation. Acquired causes can be divided into the following groups: infectious diseases (syphilis and syphilitic aortitis, bacterial endocarditis, mycotic aortitis, tuberculosis), degenerative diseases (atherosclerotic injury, cystic medial necrosis without connective tissue disease), chest trauma, vasculitides and inflammatory diseases (Bechet's disease, Takayasu arteritis, giant cell arteritis, sarcoidosis), iatrogenic diseases (valve replacement due to any cause, percutaneous manipulations in the aortic root). In many cases the cause remains unknown.

The clinical picture depends on the size and position of the aneurysm sac, anatomy of coronary arteries and their ostium, the primary cause of the lesion, and compression signs of bordering cardiac structures, but most importantly on the presence or absence of rupture. Unruptured SoV aneurysms are usually asymptomatic, therefore they are most often diagnosed accidentally (8, 9). However, unruptured SoV aneurysms may produce symptoms when they become infected or the pressure of such nearby structures as coronary arteries occurs (10). Ischaemic heart disease, including angina pectoris, is a rare manifestation of SoV aneurysm (4); however, cases of fatal ST elevation myocardial infarction have been described

(12, 13). SoV aneurysm can cause myocardial ischaemia by directly compressing coronary vessels as well as impairing haemodynamic parameters at the ostium of a coronary vessel. The authors believe that both mechanisms were present in the case presented.

On the other hand, in most cases the pathology is diagnosed after rupture occurs (5, 9). However, even ruptured aneurysm presents a diverse clinical picture: from completely asymptomatic to a cardiogenic shock and sudden death (12). RSoV aneurysm commonly protrudes and ruptures into the right ventricle, NCSoV aneurysms tend to rupture into the right atrium. LSoV aneurysms typically rupture into the pulmonary artery, the left ventricle, the myocardium or even the pericardium (4). The signs and symptoms of ruptured aneurysm are highly dependent on the size, position, and the function of the shunt.

Most commonly detected concomitant abnormalities are the ventriculoseptal defect (30–60%), bicuspid aortic valve (10–20%) (7, 10), and right ventricular outflow tract stenosis (7.5%) (14). Other valvular abnormalities, atrioseptal defect, and aortic coarcation are much rarer associated findings (15). Apart from that, SoV aneurysm itself often causes a prolapse of the corresponding aortic valve cusp that can lead to significant aortic regurgitation.

Transthoracic echocardiography is sufficient to precisely diagnose SoV aneurysms (>93.9% of cases) and its complications, including rupture (>89.2% of cases) (7, 14). The aneurysm sac was clearly visualized by means of echocardiography at our institution, however, the referring cardiologist did not describe the abnormality. The growing number of consultations and the shortening of consultation length rise medical safety concerns worldwide (16). Due to the lack of time, the quality of echocardiography is becoming lower; clinicians are unable to perform a full echo-screening and only pay attention to the most common sights that are related to angina pectoris (contraction defects and valvular abnormalities). This makes rare yet easily detectable causes of angina pectoris (such as SoV aneurysm) undiagnosed before referral to an invasive procedure.

Transesophageal echocardiography is required in up to 25% of case to better visualize the relationship of the aneurysm sac and bordering structures (7). If diagnostic doubts regarding anatomy of the lesion remain, ECG-gated cardiac CT angiography is an excellent method to better visualize the aneurysm, its relationship with bordering structures including coronary arteries, as well as to precisely describe the position of coronary artery ostium. Apart from that, it is a useful method to analyse coronary arteries for obstructive changes if invasive coronary angiography is not technically possible or the risk of the invasive procedures is too high. The advantages of ECG-gated CT angiography were therefore taken into account at our institution when planning the treatment. If echocardiography acoustic windows are inadequate and further diagnostic workup does not provide answers to the questions regarding the biventricular function and valvular pathology, or doubts still remain, cardiac MRI is a helpful non-invasive diagnostic modality (7).

There are no randomized controlled trials comparing the efficacy and safety of different SoV aneurysm treatment modalities. The majority of the data regarding treatment options and outcomes come from case reports, surgical treatment series, and single centre experiences. Few papers summarize multicentre data. Traditionally, open heart surgery has been a method of choice (8, 17, 18). If the aortic valve is intact and aortic regurgitation is insignificant, it is relevant to perform a valve-sparing surgery to avoid long-term anticoagulation and prosthetic valve-related complications (18). On the other hand, percutaneous aneurysm closure techniques have been offered as an alternative to open surgery and the results of such modalities are promising (19, 20). Long-term results of open heart surgery in the setting of SoV aneurysm are good to excellent (8), this treatment modality is considered safe and effective for both ruptured and unruptured aneurysms. It is important to point out that elective surgery has a much better prognosis than emergency intervention. The treatment method of choice depends on the cause of the aneurysm, its size, anatomy and function of the aortic valve, haemodynamics, functional status of the patient, life expectancy, desired anticoagulation strategy, and, most importantly, the experience of the surgical team (21).

CONCLUSIONS

Unruptured LSoV aneurysm-caused angina pectoris is an extremely rare medical condition. The presented case reveals the importance of

high-quality non-invasive investigation prior to coronary angiography. The majority of SoV aneurysm cases are congenital. Open heart surgery has been a treatment of choice for decades, but percutaneous aneurysm closure techniques are nowadays used in some centres with promising results. There is a high need of a randomized multicentre controlled trial with a sufficient sample size to compare the results of both treatment options. Until such a trial is carried out, the authors suggest choosing the method of treatment which the cardiovascular centre is experienced in.

CONFLICT OF INTEREST

None declared.

ETHICAL STATEMENT

This research was conducted according to the Declaration of Helsinki. Both authors have equally contributed to the article and approved the final version.

INFORMED CONSENT

Informed consent to publish the case was obtained from the patient.

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KRŪTINĖS ANGINA IR VALSALVA SINUSO ANEURIZMA: NEĮPRASTAS RETOS LIGOS PASIREIŠKIMAS

Santrauka

Valsalva sinuso aneurizma yra retai diagnozuojama aortos liga. Straipsnyje aprašomas 57 metų pacientės, kuriai diagnozuota kairiojo Valsalva sinuso aneurizmos sukelta krūtinės angina, klinikinis atvejis. Analizuojama klinikinė situacija yra sudėtinga ir išskirtinė, kadangi pacientei diagnozuota tiek Valsalva sinuso aneurizma, tiek aortos koarktacija. Liga pasireiškė jai nebūdingais simptomais ir sukėlė miokardo išemiją. Straipsnyje pateikiama naujausių mokslinių įrodymų ir įžvalgų analizė.

Raktažodžiai: krūtinės angina, Valsalva sinuso aneurizma, įgimta širdies yda, aortos koarktacija