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RUPTURED SINUS OF VALSALVA ANEURYSM: DON'T JUDGE A BOOK BY ITS COVER!

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АНЕВРИЗМАЛНА РУПТУРА НА СИНУСА НА ВАЛСАЛВА – НЕ СЪДЕТЕ ЗА КНИГАТА ПО КОРИЦАТА!

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Abstract. **Background:** Ruptured Sinus of Valsalva aneurysm (RSOV) has poor prognosis and a high mortality rate. Clinical suspicion is critical for prompt diagnosis and management. **Case Presentation:** A 33-year-old woman with no comorbidities, presented with persistent cough and recurrent lower respiratory tract infection for 2 years. Echocardiogram showed ruptured right sinus of Valsalva into right ventricle with calcified ruptured membrane forming wind-sock morphology. When the patient was taken to the operating room, it was discovered that the RSOV had ruptured into the right atrium. She underwent excision of calcified wind-sock tissue along with pericardial patch closure of ruptured right sinus. **Conclusions:** We discuss a case of an RSOV with atypical presentation, as well as a disparity in imaging features and surgical findings.

Key words: ruptured right sinus of Valsalva, wind-sock deformity, sinus of Valsalva repair, case report, surgery

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Резюме. **Въведение:** Аневрималната руптура на синуса на Валсалва (RSOV) е с лоша прогноза и висока смъртност. Клиничното подозрение е от решаващо значение за бързото диагностициране и лечение. **Представяне на случай:** Представяме случая на 33-годишна пациентка без съпътстващи заболявания, със симптоматика от упорита кашлица и рецидивираща инфекция на долните дихателни пътища в продължение на 2 години. Ехокардиограмата показва руптурирал десен синус на Валсалва в дясната камера с калцифицирана разкъсана мембрана, образуваща морфология тип „ветропоказател“¹. При превеждането на пациентката в операционната е установено, че RSOV е разкъсана в дясното предсърдие. Пациентката претърпя ексцизия на калцираната тъкан на разкъсаната мембрана и поставяне на перикарден пач за затваряне на разкъсания десен синус. **Заключение:** Представихме случай на RSOV с атипично представяне, както и с несъответствие между образните характеристики и хирургичните находки.

Key words руптура на десен синус на Валсалва, wind-sock deformity, възстановяване на синус на Валсалва, клиничен случай, хирургия

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¹Симптомът "ветропоказател" (windsock) се отнася до изяви, наблюдавани при дисекции на гръдната аорта тип А при КТ с контраст. Получава се от интимоинтимална инвагинация между истинския и фалшивия дисектиран лумен на гръдната аорта. Променящата се плътност на контраста между лумените на дисекцията, които се стесняват дистално, наподобяват ветропоказател.

BACKGROUND

A sinus of Valsalva aneurysm (SOVA) is an enlargement of the aortic root area between the aortic valve annulus and the sinotubular ridge [1]. Ruptured sinus of Valsalva aneurysm (RSOV) is rare feared complication of SOVA. The anatomical location of the aneurysm determines the consequences of rupture. The right sinus of Valsalva aneurysm commonly ruptures into right ventricle, non-coronary sinus of Valsalva aneurysm ruptures into right atrium, and the left sinus of Valsalva aneurysm ruptures into the pericardial cavity. When the RSOV communicates with the right chambers of the heart, a significant left-to-right shunt ensues, resulting in continuous murmurs, and progressively worsening heart failure [2]. Patients usually present with shortness of breath, palpitations, chest pain, fatigue, and syncope. Definitive therapy includes surgical repair or device closure as appropriate. We present the surgical management of RSOV ruptured into the right atrium with atypical presentation, and imaging features that did not correlate with surgical findings.

CASE PRESENTATION

A 33-year-old woman with no known comorbidities reported to our institution with persistent cough. She reported that she had been managed conservatively with

medications for 2 years. She had no history or physical findings suggestive of infective endocarditis, and her blood culture was negative. She had no chest pain, palpitations, and other peripheral signs of right heart failure. Family history was unremarkable. On further evaluation, her chest x ray revealed cardiomegaly, and enlarged pulmonary arteries (Fig. 1), electrocardiogram showed non-specific T wave abnormality with prolonged QT, and echocardiogram showed ruptured right sinus of Valsalva into right ventricle with calcified ruptured membrane forming wind-sock morphology, moderate tricuspid regurgitation with moderate pulmonary artery hypertension and normal left ventricular function (<https://10.3897/bgcardio.29.e109296.suppl.1> – Echocardiogram showing ruptured right sinus of Valsalva into right ventricle with calcified ruptured membrane forming wind-sock morphology, moderate tricuspid regurgitation with moderate pulmonary artery hypertension and normal left ventricular function). The computed tomography (CT) aortogram revealed that the rupture had occurred into the right ventricle, with a defect size of 5.5 mm morphology (Fig. 2A) and a 1.4 cm calcified lesion attached to the ruptured membrane of the aneurysm, resulting in a “wind-sock” morphology (Figure 2B). The patient was taken up for RSOV repair through midline sternotomy and cardiopulmonary bypass using aortic and bi-caval cannulation. Surprisingly, during surgical examination, we discovered that the RSOV had ruptured into the right

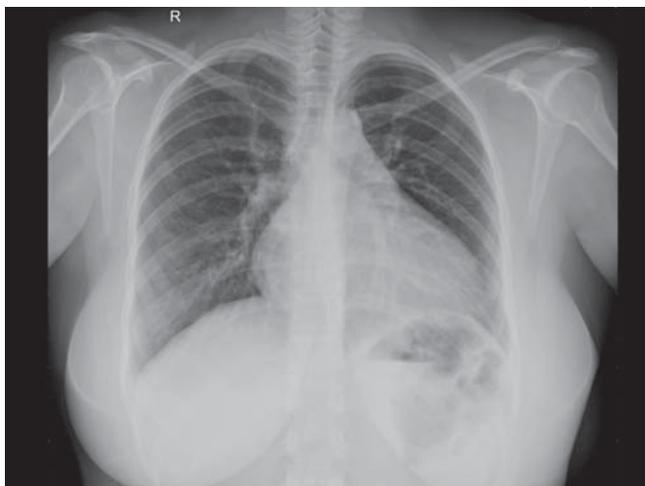


Fig. 1. Chest x ray showing cardiomegaly, and enlarged pulmonary arteries

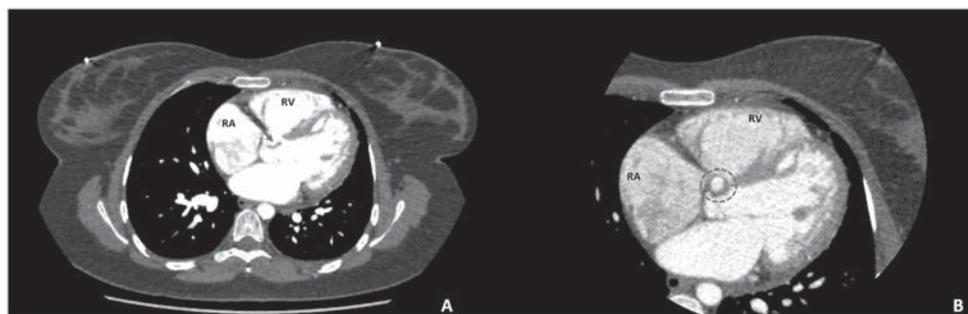


Fig. 2. Computed tomography aortogram revealing rupture of the sinus of Valsalva into the right ventricle, with a defect size of 5.5 mm morphology (Figure 2A) and a 1.4 cm calcified lesion attached to the ruptured membrane of the aneurysm, resulting in a “wind-sock” morphology (Figure 2B)

atrium rather than the right ventricle, as radiologic investigations had suggested. As a result, we proceeded to open the right atrium and remove the calcified tissue. Excised tissue specimen was sent for culture, and the ruptured sinus was repaired using autologous pericardium. The postoperative course of the patient was stable, and she was discharged on the 5th postoperative day. Her three-month follow-up echocardiogram revealed no residual defect and she reported improvement of her symptoms (<https://10.3897/bgcardio.29.e109296.suppl.2> – Echocardiogram at 3-month follow-up with no residual defect).

DISCUSSION

The aortic root bulges outwards to form the three sinuses, two of which give rise to the main coronary arteries [3]. SOVA is an abnormal dilatation of the sinus of Valsalva. The aneurysms originate predominantly from the right coronary sinus (70%), or the non-coronary sinus (25%) and are more prevalent in males and individuals of Asian descent [4]. It is thought to occur due to deficiency of elastic and muscular tissue at the junction of aortic media, and the annulus fibrosis of aortic valve [5]. The diagnosis of RSOV itself is an indication for surgery because it can rupture or compress adjacent structures at any time, and cause aortic regurgitation. Clinical presentation is highly variable. Patients may be asymptomatic or report acute symptoms such as chest pain with signs of intractable heart failure. Our patient presented with persistent cough. She had been on medications for over 2 years for her compliant. Echocardiography continues to be the most commonly employed diagnostic technique for RSOV. Cardiac CT and sine magnetic resonance imaging are increasingly being used to visualize the precise cardiac anatomy [6]. It's worth noting that the echocardiography and aortogram both suggested an RSOV aneurysm rupturing into the right ventricle. The tissue from the aneurysmal sac protruding into the atrium was not obvious in the radiologic imaging probably because of the windsock effect. Hence, it was thought to be protruding into the right ventricle which is a common presentation. During surgery, there was no rupture into the right ventricle. We opened both the aorta and the right atrium and found calcified tissue in the right atrium. The

tract was excised, and a pericardial patch was used to close the aortic side of the opening. Several reports of RSOV rupture into the right atrium have surfaced in the recent times [6-8]. The uniqueness of report is that the patient's complaints of persistent cough which was suspected to a recurrent respiratory infection and treated with medications for over 2 years, combined with the fact that imaging characteristics did not match surgical findings. Clinical suspicion is the key to correct diagnosis. It is well known that radiologic investigations help guide therapy. Clinical judgement is important to address situations where radiologic findings may not match what we perceive on table.

CONCLUSIONS

Ruptured sinus of Valsalva is a rare but fatal cardiac surgical emergency which needs early intervention to prevent morbidity and mortality.

No conflict of interest was declared

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