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

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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-scock 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-scock 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-scock deformity, sinus of Valsalva repair, case report, surgery

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Background: The technical details of the patient's presentation and management are discussed in the context of the literature. The case report highlights the importance of prompt diagnosis and management of RSOV, as well as the challenges of atypical presentations and imaging features.

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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 inves-
tigations 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 pericardi-
um. 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 com-
press adjacent structures at any time, and cause aortic
regurgitation. Clinical presentation is highly variable.
Patients may be asymptomatic or report acute symp-
toms 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
complaint. Echocardiography continues to be the most
commonly employed diagnostic technique for RSOV.
Cardiac CT and cine magnetic resonance imaging are
increasingly being used to visualize the precise cardiac
anatomy [6]. It’s worth noting that the echocardiogra-
phy and aortogram both suggested an RSOV aneu-
rysm 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 pro-
truding into the right ventricle which is a common pre-
sentation. 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 cardi-
ac surgical emergency which needs early intervention
to prevent morbidity and mortality.

*No conflict of interest was declared*

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