

Case report



## CASE OF A FEMALE PATIENT WITH ANKYLOSING SPONDYLITIS AND MULTIPLE ANEURYSMS

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### ABSTRACT:

Aneurysms of the Valsalva sinuses are rare among cardiovascular disorders, with an incidence rate of 0.15%. Most cases are accidentally diagnosed or constated post-mortem. Usually, this type of pathology is asymptomatic and might go undetected for a long time before the aneurism erupts. Patients with larger coronary aneurysms often complain of from chest pain, shortness of breath, and arrhythmia. Another extremely rare cardiovascular pathology is the aneurysm of the subclavian artery, which has a reported rate of 0.13%. Smaller subclavian aneurysms have no clinical manifestation, whereas larger ones cause murmur, pulsation phenomenons, neuropathic pain and ischemia of the upper limb due to compression of the brachial plexus and major vessels.

Herein, we report a rare case of a very large aneurisms involving all three coronary sinuses and the whole aortic root in combination with a bilateral giant subclavian aneurism. These findings were made during a routine cardiological examination and further hospitalizations of a 54-year-old woman with chest pain and high blood pressure complaints with known ankylosing spondylitis. Despite their rarity, aneurysms of the sinuses of Valsalva and subclavian arteries are of utmost clinical significance due to the life-threatening complications which might arise from their rupture. Furthermore, such aneurysms pose significant technical difficulties during angiographic procedures. Therefore, detailed knowledge of such pathologies is essential for correctly diagnosing and treating such patients.

**Keywords:** Aneurysm, Aortic, Coronary, Ankylosing spondylitis, Cardiovascular,

### BACKGROUND

Sinus of Valsalva aneurysms (SVAs) is a rare cardiovascular pathology with an incidence rate that ranges between 0.09% (smith) in the autopsy series and 0.15% [1] in living patients undergoing cardiac surgery. They are more commonly found in men and Asians [2] and most often involve the noncoronary sinus. In the vast majority of cases, SVAs do not exert any clinical symptoms before they erupt. Only larger SVAs are associated with chest pain, shortness of breath and conduction disorders [3, 4].

Subclavian artery aneurysms (SAAs) are rarely encountered, with a reported incidence rate of 0.13% [5]. They most commonly occur due to atherosclerosis, trauma, or post-stenotic dilated aneurysms caused by thoracic outlet syndrome. Less frequently, infective or syphilitic media necrosis can also be responsible [6]. The symptoms of a subclavian artery aneurysm depend on its location and size. An extrathoracic aneurysm typically presents as a pulsatile lump above the clavicle, accompanied by pulsation and vascular murmurs. On the other hand, an intrathoracic aneurysm or post-stenotic dilated aneurysm can compress the brachial plexus or blood vessels in the upper extremity, leading to limb ischemia. If the aneurysm erodes the lung apex, it may cause hemoptysis, and compression of the recurrent laryngeal nerve can result in a hoarse voice. Some cases have also reported dysphagia and Horner’s syndrome [7].

Even though SVAs and SAAs are extremely rare, detailed knowledge of their clinical manifestation, diagnosis and treatment is paramount for cardiologists and cardio surgeons. This article aims to report a rare case of a giant SVA combined with bilateral SAAs in a patient.

## CASE DESCRIPTION

A 54-year-old female was admitted to our department with high blood pressure, shortness of breath, swollen legs, difficulty in swallowing and complaints of angina pectoris for several days. Her cardiovascular risk factors were high blood pressure, dyslipidemia and smoking. The patient was diagnosed 7 years ago with ankylosing spondylitis. From the physical examination we have a data for impaired general condition with kyphoscoliotic thorax, systolic murmur with punctum maximum on Erb's point and swelling of the lower limbs. We observed also stiffness in the joints. The ECG shows sinus rhythm, 71 bpm and left anterior fascicular block. The following echocardiography reviewed dilatation of the noncoronary aortic sinus of Valsalva – 68 mm; sinotubular junction – 58mm; descending aorta 38mm in diameter; left atrium 47/57 mm; right atrium – 38/49 mm; left ventricle – preserved segment kinetics. End-diastolic diameter/end-systolic diameter – 57/26 mm; end-diastolic volume/end-systolic volume – 104/38 ml; ejection fraction – 64%. Aortic valve – second-grade regurgitation; mitral valve – first-grade regurgitation; tricuspid valve - first-grade regurgitation. The blood tests showed elevated total cholesterol and LDL. The cardio-specific blood markers Troponin (hs), CPK and MB were normal. Due to the patient's elevated cardiovascular risk and the echocardiographic findings, a CT coronarography with aortography was performed. The most reviewed massive dilatation of the aortic root at the level of the Valsalva sinuses – 85x72 mm. The noncoronary sinus was the most dilated one – 60 mm; the right coronary sinus was 35 mm, and the left was 31 mm. sinotubular junction – 31,5x25 mm. Ascending aorta, at the level of the pulmonary trunk – 33x32 mm with calcium plaques. Aortic arch – 28x26 mm with calcium plaques. Brachiocephalic trunk – contrasted without significant stenosis or dilatation. Right subclavian artery – dilated with fusiform aneurism 53mm long and 46x41mm wide. (Fig.2, Fig.3 and Fig 4.) Both thrombosis and calcification can be observed on the aneurismal wall. The distal segment of the subclavian artery, after the aneurism, and the proximal segment of the axillary artery are visualised with diffuse atheromatous changes, mixed plaques and irregular lumen without data for any significant stenosis. Left subclavian artery – initially angled with a 4,5 mm stenosis in the area of the angulation, followed by a fusiform dilatation with the size of 31x27 mm, thrombosis and calcification on the wall. Distally the fusiform dilatation continues for 13 mm. Common carotids and vertebral arteries – normal contracture, without data for stenosis or dilatations. Both vertebral arteries arise from the dilatated aneurismal parts of the subclavian arteries.

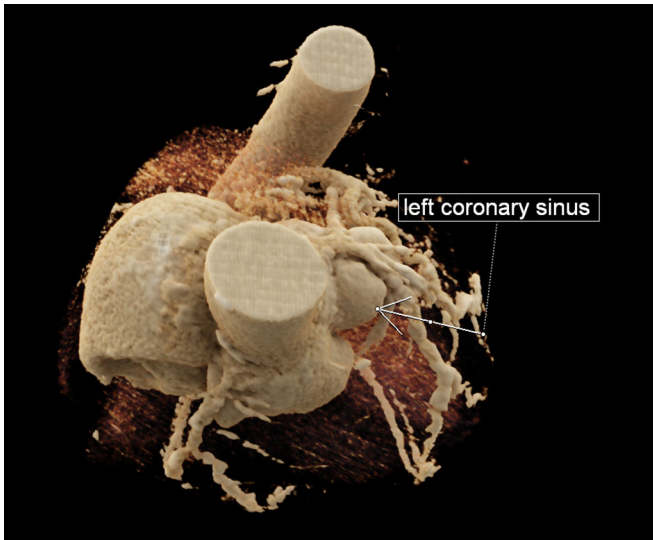
**Fig. 1.** 3D computed tomographic reconstruction of the heart (front view) showing the extremely dilated coronary sinuses. The coronary vessels that originate from them are traced with pronounced atherosclerosis.



**Fig. 2.** 3D computed tomography reconstruction of the heart (profile projection) showing the extremely dilated noncoronary sinus. Its uneven anterior contour is due to incomplete parietal thrombosis.



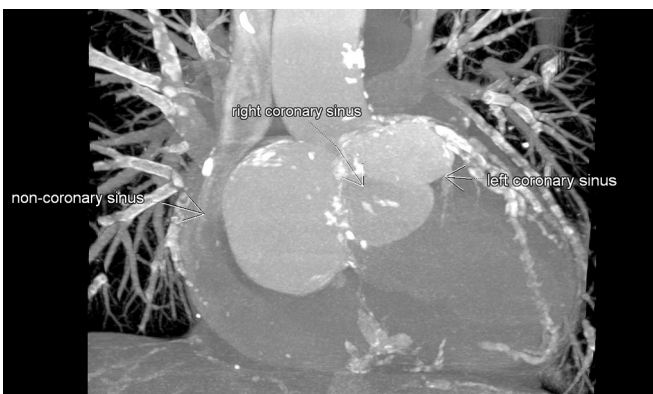
**Fig. 3.** 3D computed tomography reconstruction of the heart (top view) with annotation of the left coronary sinus.



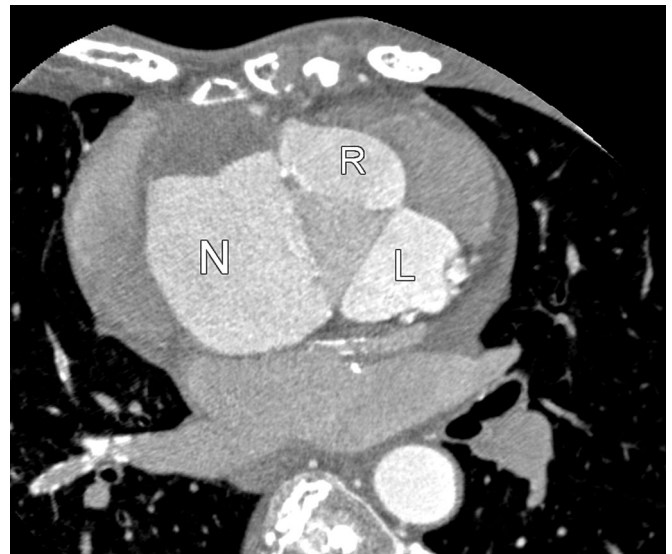
**Fig. 4.** 3D computed tomography reconstruction of the heart (bottom view) with noncoronary sinus annotation.



**Fig. 5.** Maximum intensity projection reconstruction in front view showing the dilated coronary sinuses and the significant vascular calcinosis.



**Fig. 6.** Axial image of the CT coronary angiography at the level of the aortic valve, on which the extremely dilated coronary sinuses are indicated (L – left; R – right; N – noncoronary sinus). The ventral surface of the noncoronary sinus has a parietal thrombosis.



Furthermore, coronary angiography was performed for better visualization of the coronary arteries in order to obtain a better understanding of the perfusion of the myocardium. A right trans-radial approach with a 5F JL 3,5 catheter was performed. After successful catheterization of the LAD, the following was reviewed: the main part appeared normal, LAD – 80% stenosis; LCx – 80% stenosis. The RCA had several plaques alongside the artery without data for significant stenosis. We contemplated stenting the stenotic parts; however, ultimately, we decided against it due to the high risk of failure and potentially life-threatening complications. The patient was given medical therapy and directed to consult with a cardio surgeon. Currently, the patient was rejected for surgical treatment by Heart Team from multiple hospitals due to the extremely high cardiovascular risk. The patient was left on a medical therapy which includes Bisoprolol 10 mg per day, Candesartan 16 mg per day, Dapaglifozine 10 mg per day, Torasemide 10 mg per day, Rosuvastatin 20 mg per day and Clopidogrel 75 mg per day. We recommend echocardiographic follow up on every 6 months and Holter ECG every year.

## DISCUSSION

We present a rare case of a 54-year-old female very large aneurysms involving all three coronary sinuses and the whole aortic root in combination with a bilateral giant subclavian aneurysm. Sinus of Valsalva aneurysms are an

extremely rare cardiac pathology first reported in 1839 by Hope [8]. One year later, Thurnam described the clinical manifestation of SVAs and noted the rarity of this pathology [9]. Since after several studies aiming to identify the incidence rate of SVAs were conducted. Smith reported a 0.09% incidence rate of SVAs out of 8138 autopsy series [10]. According to Takach TJ, et al., SVAs are found in 0.15% of patients undergoing cardiac surgery [1]. According to Moustafa S, et al., only 5% of all SVAs involve the left coronary sinus, 70% involve the right coronary sinus, and 25% involve the noncoronary sinus. [2] In our case, however, all three coronary sinuses are involved to different extents, as the noncoronary one is mainly involved. In contrast, giant aneurism involves the left coronary sinus the least. Clinical manifestation of SVAs involves a wide range of symptoms and is often associated with aneurysmal rupture. Nevertheless, SVAs might compress the adjacent coronary artery depending on their size, leading to myocardial ischemia. Right-sided aneurysms might compress the right atrium and cause different conduction disorders [3]. Regardless of their size, all SVAs might lead to life-threatening complications if ruptured. Thus, correct diagnosis and management of such rare cases are paramount for the patient's outcome [11].

Moreover, SVAs might lead to several complications. Aortic regurgitation is one such complication found in 30-50% of patients with SVA [3]. Nonruptured SVAs might interfere with the function of the mitral or tricuspid valve, depending on which side they propagate, which might lead to mitral or tricuspid regurgitation and exert the pathological organic murmur [4]. In the vast majority of cases, SVAs rupture into the right ventricle and quickly cause acute heart failure. According to Bricker et al., 55.6% of SVAs rupture toward the right ventricle, 30.3 into the right atrium, 8.5% into the right ventricular outflow tract, 2.1% into the left ventricle, 1.6% into the interventricular septum and 1.1% into the left atrium [12]. In addition, SVAs might rupture outwards into the extracardiac space, which is associated with a higher mortality rate due to the development of cardiac tamponade [11]. Management of such pathology requires a multidisciplinary approach. 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease: recommends surgical repair in patients with sporadic aneurysms larger than 5.5 cm and larger than 50 cm in individuals with bicuspid valves with risk factors and larger than 4.5 cm in the presence of connective tissue disease. Repair of SVA should be considered if the aneurysm shows a growth rate of more than 0.5 cm/year. The mortality rate for surgical repair ranges from 1.9% to 3.6%. Survival rates after 15 years are approximately 90% [13].

Sinus of Valsalva aneurysms are another very rare cardiac pathology. According to Dent et al., SAAs were present in only 2 out of 1488 cases, which computes an

incidence rate of 0.13% [5]. Clinical manifestation of SAAs involves a broad amount of symptoms determined by the localization and size of the aneurysms. Intrathoracic aneurysms present a plethora of symptoms resulting from the compression of adjacent structures. Such symptoms include

- Horner's syndrome, the result of compression of the sympathetic trunk;
- hoarse voice due to compression of the recurrent laryngeal nerve;
- Paresthesia, neuropathic disorders, and superior limb ischemia are caused by brachial plexus and major vessels compression [7].
- Seldom dyspnea has been reported as a symptom as a consequence of tracheal compression caused by a gigantic SAA [14].

Extrathoracic SAAs manifest with pulsation and vascular phenomenon localized in the supraclavicular fossa [8]. Moreover, SAAs are associated with another type of aneurysms in 30% to 47% of cases, as aortic ones are the most common [7, 15, 16]. Therefore, patients with SAAs should be thoroughly checked for other aneurysms. Furthermore, SAAs are associated with a significant risk of dissection, thrombosis and/or rupture, which might result in life-threatening complications. In this case, surgical correction of SAAs is strongly advised [17]. The surgical treatment for subclavian artery aneurysms involves resecting part of the clavicle or sternum, which poses risks such as injury to adjacent vessels or the brachial plexus [Vierhout]. The development of endovascular techniques has led to the use of less invasive stent-graft insertion, but the subclavian artery presents challenges like endoleak and potential stent kinking or fracture [18, 19]. Endoleak is a common complication requiring regular follow-up. [19, 20] In cases of an enlarged aneurysmal sac and mass effect symptoms, surgical removal of the aneurysm is preferred. Open surgical repair aims to remove the aneurysm and maintain blood flow continuity but may require clavicular resection or sternotomy. Treatment choice depends on the aneurysm's location and size, and continuous follow-up is crucial for both endovascular repair and open surgery [21]. There is also a relation between ankylosing spondylitis and cardiovascular diseases. Inflammatory conditions are a rare cause of aortic aneurysms, accounting for 3% to 10% of cases. Patients with ankylosing spondylitis uncommonly present with ascending aortic aneurysms related to long-standing, aggressive disease. Miller et al. reviewed the case of a young man with ankylosing spondylitis exhibiting complex inflammatory aortic aneurysms atypically involving the abdominal and descending thoracic aorta, as well as ectasia of medium-sized visceral vessels. [22].

SVAs and SAAs have meagre incidence rates on their own, to begin with; however, in our case, we have a combination of a gigantic SVA involving the whole aortic root

and all three coronary sinuses in combination with bilateral SAAs. Very few similar reports can be found throughout the literature, none entirely the same as ours. Tanaka et al. reported a case of bilateral saccular SAAs in combination with a saccular aneurism of the aortic arch and fusiform aneurism of the infrarenal part of the abdominal aorta [22]. However, in their case, there is no SVA nor any involvement of the coronary sinuses reported.

## CONCLUSION

Encountering patients with SVAs and/or SAAs is a rare yet plausible scenario. This type of cardiovascular pathology might be asymptomatic or present with vague symptoms. For that reason, detailed knowledge of these pathologies is of utmost clinical significance for the correct diagnosis and management of such patients. This pathology should be observed in case of present connective tissue disorder with pathological murmors from the physical examination. The diagnosis requires a global exploration of the arterial bloodflow with CT or MRT imaging.

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