

A Giant Coronary Sinus Aneurysm with Secondary Vena Cava Aneurysms

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ABSTRACT

Coronary sinus aneurysm (CSA) is an extremely rare entity. Herein, we present an unusual case of an 18-year-old symptomatic female patient with a giant CSA. Secondary vena cava aneurysms were also manifested. The final diagnosis was confirmed by enhanced computed tomography (CT) and cardiac catheterization. As far as we know, it is the first case that such a giant CSA coexists with secondary vena cava aneurysms. Considering the complexity of postoperative reconstruction, we believe that heart transplantation may be the optimal way for treatment. The patient received anticoagulant due to the superior vena cava (SVC) thrombosis while waiting for a donor.

INTRODUCTION

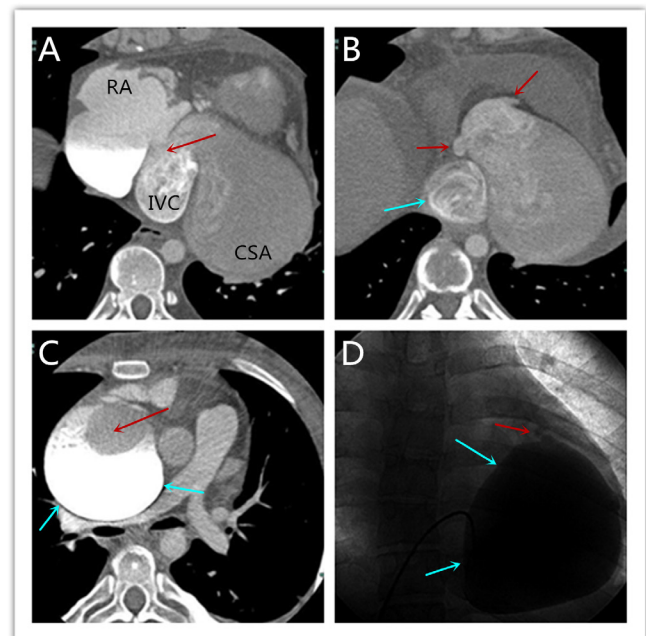
There are many reports about the anomaly of coronary sinus, however, less than 100 surgically treated cases have been reported in English literature. Coronary sinus aneurysm (CSA) is primarily induced by coronary artery fistula, while congenital cardiac malformation constitutes a few cases [Vijay 2014]. The treatment of CSA still remains controversial. Meanwhile, the vena cava aneurysm is equally rare and only a few cases have been reported [Davidovic 2008]. We present a unique case of CSA coexisting with secondary vena cava aneurysms. Considering the risk and difficulty of the corrective surgery, heart transplantation was suggested.

CASE REPORT

An 18-year-old female was admitted with a history of chest distress as well as recurrent pitting edema for more than 15 years. She underwent a pericardial drainage because of heart failure eight years ago, which was diagnosed as constrictive pericarditis. In recent years, the symptoms of heart failure were worsening. Abdominal ultrasound revealed hepatomegaly and moderate ascites. Electrocardiograph showed sinus rhythm and occasional atrial premature beats. Transthoracic echocardiography (TTE) revealed a giant cystic lesion (7.5 × 5.7 × 7.3 cm) posterior to the wall of the left heart. Enhanced computed tomography (CT) demonstrated a giant cystic lesion (9.7 × 7.2 cm) connecting to the junction between right

atrium and inferior vena cava (IVC) (Figure, A), which was diagnosed as CSA because three coronary veins draining into it were observed (Figure, B). Moreover, both superior vena cava (SVC) and inferior vena cava (IVC) aneurysms with SVC thrombosis were also manifested (Figure, B and C). In addition, enhanced CT suggested the irregular and hypoplasia right heart. For further assessment, right heart catheterization was performed and clarified the relationship between coronary veins and CSA (Figure, D), which elucidated the CT's finding. All above clinical manifestation and imaging results indicated the final diagnosis: concomitant congenital CSA, secondary vena cava aneurysms, and SVC thrombosis.

After careful consideration, we preferred heart transplantation for this patient. During the waiting period, diuretic and anticoagulant therapies were used for improvement of heart function and prevention of further thrombosis, respectively. The patient has been followed up for six months without deterioration.



A, Enhanced CT showed the CSA originating from the connection (red arrow) between RA and IVC. B, Enhanced CT revealed two coronary veins (red arrows) draining into CSA and IVC aneurysm (blue arrow). C, Enhanced CT showed the SVC aneurysm (blue arrows) and intravascular thrombosis (red arrow). D, Cardiac catheterization showed a giant cystic lesion (blue arrows) which was diagnosed as CSA due to the detection of drainage from the coronary vein (red arrow).

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DISCUSSION

CSA is an extremely rare disease with less than 100 cases being surgically treated. Embryologically, coronary sinus is derived from the left horn of sinus venosus (eighth week of gestation) [Igawa 2006]. In this case, without evidence of coronary artery fistula, the CSA could be a remnant of the left horn of sinus venosus, which may develop in the gestational process of formation of coronary sinus.

The clinical manifestation of CSA depends on multiple factors such as age, aneurysm size, and individual tolerance. According to Binder's study, CSA is usually symptomatic and most commonly presents with symptoms including arrhythmia, palpitation, chest discomfort, shortness of breath, and fatigue [Binder 2000].

Electrocardiography of patients with CSA usually shows sinus rhythm, atrial fibrillation/flutter, or incessant supraventricular tachycardia. In our case, TTE was useful for detection of the giant cyst but it was hard to find its origination. Enhanced CT and catheterization here were much more valuable to determine the origination of aneurysm, by showing the clear connection between the coronary veins and CSA. Additionally, they also elucidated that there was no coronary artery fistula, which was believed to be the most common reason for CSA.

In this case the giant CSA caused significant mass effect, which led to secondary changes like distortion of right heart and aneurysmal dilatation of both vena cava. The compressed right heart maintained a relatively high venous pressure, which contributed to the blood retention and compensatory dilation of vena cava. In addition, SVC thrombosis is mainly induced by slow blood flow and coagulopathy.

It is difficult to assess the prognosis of patients with such cardiac malformations since experience is not available in

many instances. There is consensus about catheter or surgical ablation in patients suffering from arrhythmias [Binder 2000]. Surgical treatment of such a giant aneurysm is clearly indicated to prevent rupture. However, simultaneous repair of the CSA and both vena cava aneurysms seemed to be impossible because of the complexity of postoperative reconstruction and the poor condition of the patient. What's worse, the previous pericardial drainage might have led to tissue adhesion and higher risk of intraoperative hemorrhage. Hence, heart transplantation was thought to be the optimal choice for the patient.

Conclusion

CSA and vena cava aneurysms are both rare entities. This case report describes a patient with the rare condition of having both disorders, which, to our knowledge, has not been reported before. CT and catheterization are helpful for final diagnosis. Due to the complexity, we believed that heart transplantation was the optimal option for this patient and anticoagulant was applied for prevention of further vena cava and coronary sinus thrombosis.

REFERENCES

- Binder TM, Rosenhek R, Frank H, Gwechenberger M, Maurer G, Baumgartner H. 2000. Congenital malformations of the right atrium and the coronary sinus. *Chest* 117:1740-8.
- Davidovic L, Dragas M, Bozic V, Takac D. 2008. Aneurysm of the inferior vena cava: case report and review of the literature. *Phlebology* 23:184-8.
- Igawa O, Adachi M, Inoue Y, Hisatome I. 2006. Giant aneurysm of the coronary sinus. *J Cardiovasc Electrophysiol* 17:690.
- Vijay SK, Tiwari BC, Misra M, Joshi LM, Srivastava DK. 2014. Aneurysmally dilated giant coronary sinus mimicking accessory cardiac chamber. *Echocardiography* 31:E70-1.