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Cor triatriatum: A rare congenital cardiac disease in differential diagnosis

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ABSTRACT

Cor triatriatum is a rare congenital cardiac anomaly which requires an urgent surgical repair. Herein, we report an infant with the diagnosis of cor triatriatum sinistrum and to highlight the importance of early differential diagnosis in congenital heart diseases.

Keywords: Congenital, cor triatriatum sinistrum, heart disease.

Congenital heart diseases can be difficult to diagnose in infants, as they often present with non-specific symptoms. Cor triatriatum is among the rarest of all congenital cardiac anomalies (0.1 to 0.4% of all congenital cardiac malformations) and is often a hemodynamically mild incidental finding.^[1]

Herein, we report an infant with the diagnosis of cor triatriatum sinistrum and to highlight the importance of early differential diagnosis in congenital heart diseases.

CASE REPORT

A four-month-old female infant with cough, rhinorrhea, and mild dyspnea was admitted to the emergency department and diagnosed with a viral illness. Chest radiograph showed moderate cardiomegaly and mild prominence of pulmonary vasculature. She was referred to the pediatric cardiology department. After examination, electrocardiogram showed sinus tachycardia with right axial deviation and right ventricular hypertrophy. Echocardiogram revealed the diagnosis of cor triatriatum sinistrum. An urgent surgical repair was decided (Figure 1). A written informed consent was obtained from each parent.

The surgical procedure included resection of the membrane at the left atrial cavity. A right atrial approach was preferred and cardiopulmonary bypass with mild-to-moderate hypothermia was applied. The early postoperative period was favorable and

transthoracic echocardiography showed the complete removal of the left intra-atrial membrane. The postoperative course was uneventful and the patient was discharged five days after the operation.

DISCUSSION

Cor triatriatum is a rare congenital cardiac anomaly which affects about 0.1 to 0.4% of all cases with congenital heart disease. First described by Church in 1868, it was described as an additional fibromuscular membrane within the left atrium on autopsy and, then, the entity was specifically named and described in detail by Borst in 1905.^[2]

A fibromuscular septum divides the left atrium into two chambers and is thought to arise from failed resorption of the common pulmonary vein. The additional membrane can occur within the left atrium (cor triatriatum sinistrum) or, much more rarely, within the right atrium (cor triatriatum dexter). Cor triatriatum is associated with additional congenital heart lesions in 80% of cases, most commonly atrial septal defects and anomalous pulmonary venous return.^[3]

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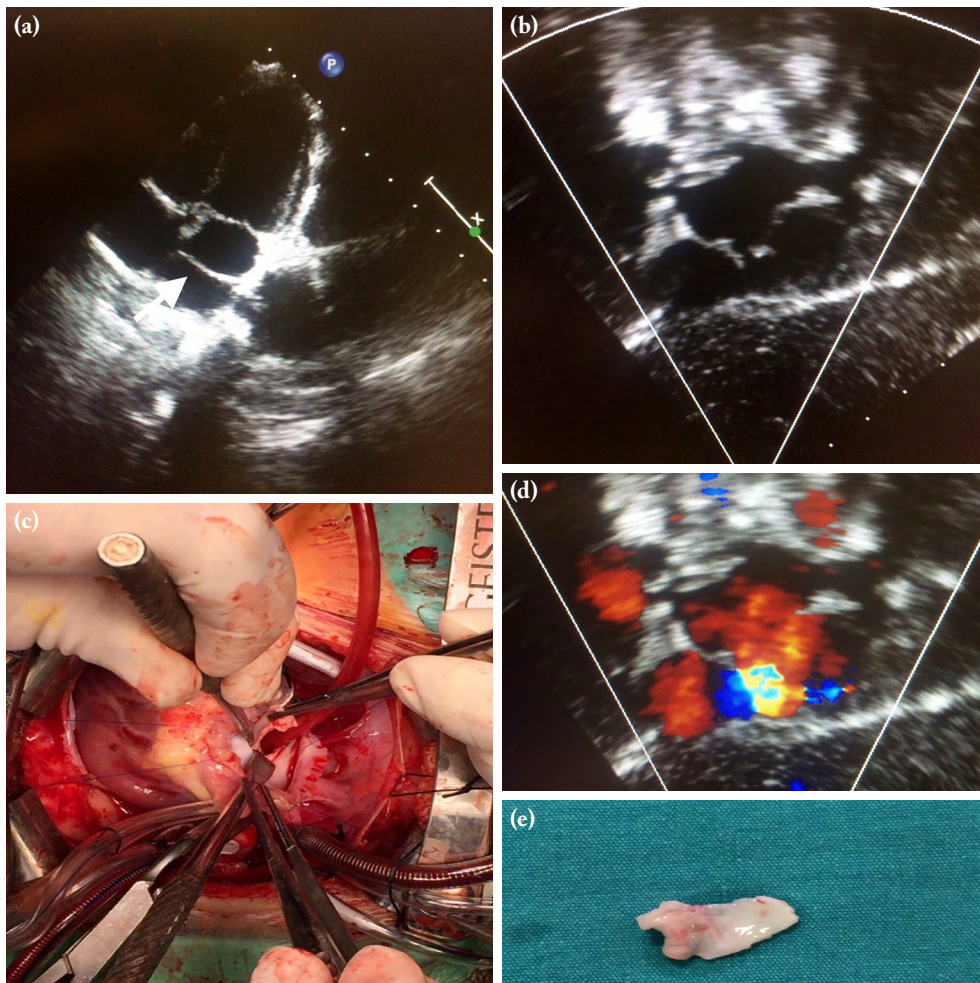


Figure 1. (a, b, d) Echocardiographic and (c) intraoperative images of cor triatriatum sinistrum and (e) image of membrane resected.

The symptom spectrum of the disease correlates with the degree of obstruction caused by the membrane. Larger openings and normal venous return correspond to fewer symptoms. Patients with a significant obstruction are likely to present in infancy with symptoms resulting from pulmonary congestion and pulmonary arterial hypertension. Common presentations include failure to thrive, dyspnea, cyanosis, or even shock.^[4] Late presentation in late adulthood may be due to fibrosis and calcification of the orifice. A long-standing turbulent flow through the membrane can cause stenosis or with the development of mitral regurgitation or atrial fibrillation. The symptoms are similar to the symptoms of mitral stenosis.^[5]

There are various techniques available to identify the pathology. As in our case, echocardiography

is diagnostic and can differentiate from other congenital heart lesions, such as pulmonary vein stenosis. More invasive diagnostic studies such as magnetic resonance imaging, computed tomography, and cardiac catheterization can be also used, when echocardiography yields uncertain findings.^[6]

Recommended treatment depends on symptoms. Mild dyspnea in older patients may improve with diuretics and preload reduction. For those with worsening or severe symptoms, as in our case, surgical correction is often required. Resection of the membrane can be curative with a rare recurrence of symptoms; however, surgical repair appears to have a greater mortality at younger ages (<5 years) and those with severe preoperative heart failure.^[7]

In conclusion, cor triatriatum is a congenital heart disease which usually presents during infancy with failure to thrive mimicking respiratory infections and sepsis. It can be life-threatening and must be rapidly diagnosed. Surgical treatment is often life-saving, even after the development of respiratory failure and shock.

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REFERENCES

1. Briasoulis A, Sharma S, Afonso L. A three-dimensional echocardiographic approach to cor triatriatum. *Int J Cardiol* 2015;180:262-3.
2. Chen Q, Guhathakurta S, Vadalapali G, Nalladaru Z, Easthope RN, Sharma AK. Cor triatriatum in adults: three new cases and a brief review. *Tex Heart Inst J* 1999;26:206-10.
3. Humpl T, Reineker K, Manlhiot C, Dipchand AI, Coles JG, McCrindle BW. Cor triatriatum sinistrum in childhood. A single institution's experience. *Can J Cardiol* 2010;26:371-6.
4. McKeag NA, Murphy JC, Dixon LJ. An incidental finding or an unusual cause for a transient ischaemic attack? *QJM* 2012;105:789-90.
5. Erden EÇ, Erden İ, Kayapınar O. Cortriatriatum sinister with significant pressure gradient in an adult patient. *Turk Gogus Kalp Dama* 2013;21:143-5.
6. Masui T, Seelos KC, Kersting-Sommerhoff BA, Higgins CB. Abnormalities of the pulmonary veins: evaluation with MR imaging and comparison with cardiac angiography and echocardiography. *Radiology* 1991;181:645-9.
7. Saxena P, Burkhart HM, Schaff HV, Daly R, Joyce LD, Dearani JA. Surgical repair of cor triatriatum sinister: the Mayo Clinic 50-year experience. *Ann Thorac Surg* 2014;97:1659-63.

Penetrating cardiac trauma mimicking congenital sinus Valsalva aneurysm rupture: A case report

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ABSTRACT

Penetrating cardiac injuries are rare, but often fatal. They may present with different manifestations. A 30-year-old male patient was admitted with self-inflicted left parasternal stab wound in the second intercostal space. Computed tomography revealed pericardial and pleural effusion. Echocardiography and aortography showed severe aortic regurgitation and aorto-right ventricular communication mimicking a congenital sinus Valsalva aneurysm rupture. The patient was operated and aortic leaflet repair with a pericardial patch and closure of the communication on both aortic and right ventricular sides were performed. In conclusion, although such traumatic injuries are rare, they may be life-threatening and, therefore, requires prompt treatment.

Keywords: Congenital sinus; heart injuries; penetrating injuries; Valsalva aneurysm rupture.

Penetrating cardiac injuries are rare, but represent a high mortality in most cases. The injury is usually non-accidental, predominantly sustained by males, and the mortality rate is about 60%, which further increases where pericardial tamponade is absent.^[1,2] In a recent large series, 86% of injuries constituted single chamber injury, predominantly right ventricle (46%) and left ventricle (24%), whereas multi-chamber injuries were seen in 18% of cases. Patients typically manifest with signs of cardiac tamponade, cardiac arrest, or signs of extremis from exsanguination.^[2]

Congenital sinus Valsalva aneurysms are rare abnormalities often presenting with rupture. In more than 80% of cases, the rupture originates from the right coronary sinus (RCS). Aorto-right ventricular connection is observed in 65% of cases.^[3,4]

Herein, we report a case of aorto-right ventricular communication concomitant with aortic regurgitation due to self-inflicted stab wound injury.

He was examined in a rural medical center and the skin wound was sutured. The patient reported no complaint subsequently. Echocardiography and aortography revealed aortic valve regurgitation due to a defect in the right coronary cusp (RCC) and aorto-right ventricular communication (Video 1). Pericardial and bilateral pleural effusion was noted on echocardiography and computed tomography (CT) scan (Figure 1a). He had no signs of tamponade or pathological findings, except for the sutured wound (Figure 1b).

He was scheduled for an urgent operation and a written informed consent was obtained. A median sternotomy was performed. Due to the possibility of aortic injury, right femoral artery was cannulated to initiate cardiopulmonary bypass (CPB) immediately following pericardiotomy. Hematoma and defibrinated blood were present in the pericardial space. A very little adventitial hematoma was located in the left lateral side of the aorta above the sinotubular

CASE REPORT

A 30-year-old male patient was admitted to the cardiology department with chest pain. He had a self-inflicted left parasternal stab wound in the second intercostal space two days ago after heavy alcohol intake. He did not remember the type of knife used.

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junction. The right atrium was cannulated and CPB was initiated. Transverse aortotomy was performed. There was a slit-like linear opening on RCS connected with right ventricular cavity (Figure 1c). There was also a 2×1 cm defect in the RCC. Transverse right ventriculotomy was performed to visualize the opening of the defect and a slit-like linear opening was noted (Figure 1d).

The aortic side of the opening was sutured with two Teflon pledgeted sutures (Figure 1e) and the right ventricular side with the Teflon felt on both sides (Figure 1f). The defect in the RCC was repaired with a fresh pericardial patch. Aortotomy was closed and the patient was weaned

from CPB. The postoperative course was uneventful and postoperative echocardiography revealed no aortic regurgitation or aorto-right ventricular communication. In the first year of follow-up, the patient is still stable with normal echocardiographic findings.

DISCUSSION

Penetrating cardiac injury is often fatal with a mortality rate over 60%. Multi-chamber injuries constitute a lower percentage of cases with a predominant single right ventricle injury.^[1,2] The present case is unique, as it is a late admission and had no signs of hemodynamic compromise,

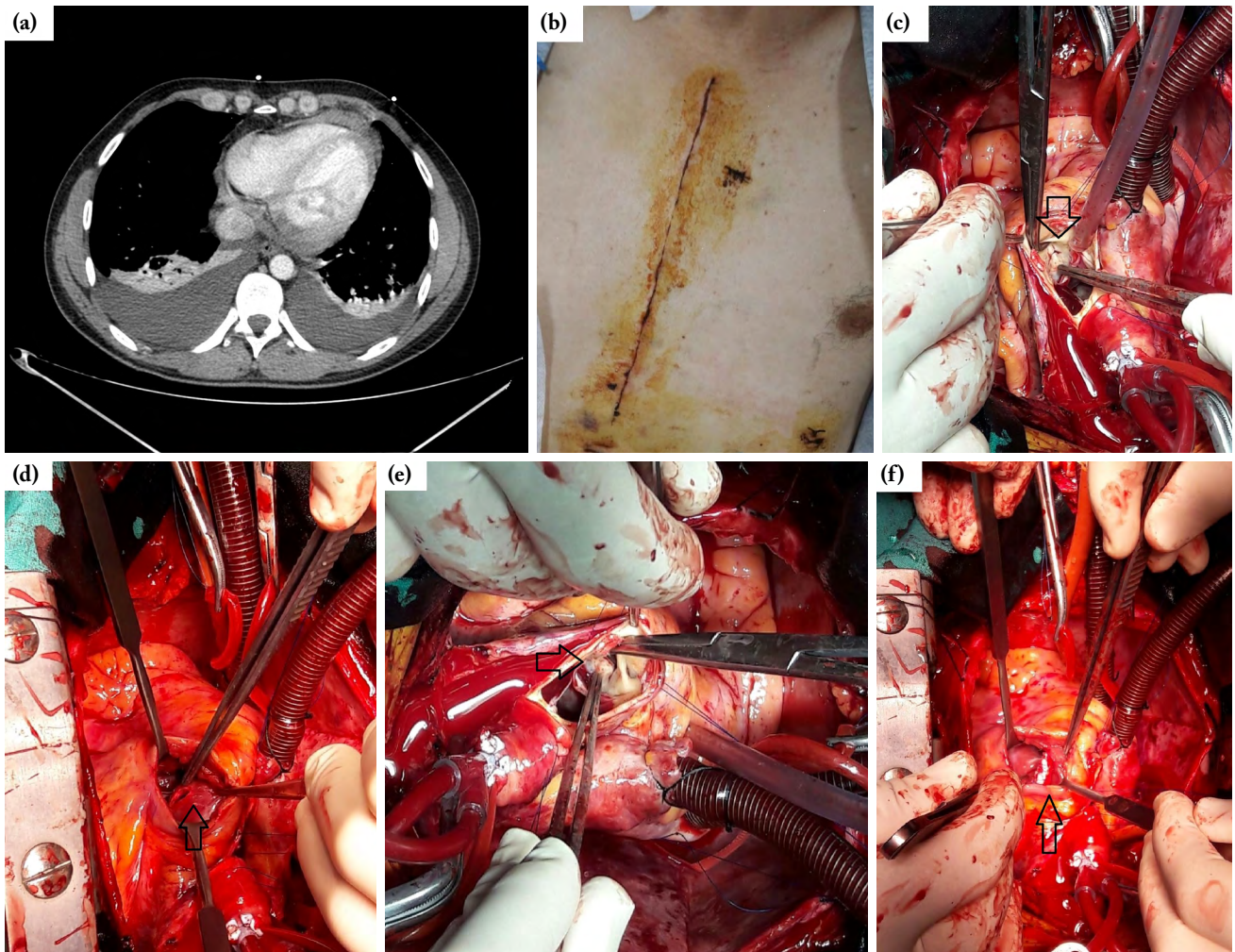


Figure 1. (a) Computed tomography scan showing pericardial and bilateral pleural effusion. (b) The stab wound injury in the second left parasternal intercostal space. (c) Aortic slit-like linear injury (arrow). (d) Right ventricular slit-like linear injury (arrow). (e) Aortic side of repair with two Teflon pledgeted sutures (arrow). (f) Right ventricular side of repair with over and over suture supported with Teflon felts on both sides (arrow).

despite a great injury with an aorto-right ventricular communication.

Such an injury might have caused a great hemodynamic compromise due to signs of exsanguination or cardiac tamponade. However, our case had no complaints at the time of injury and was admitted two days later due to chest pain. Surprisingly, pericardial effusion was not much and did not cause tamponade within two days. There was a small adventitial hematoma in the left lateral side of the aorta, however, there was neither hemorrhage from the aorta nor a visible aortic injury site. In this case, the injury mimicked a ruptured congenital sinus Valsalva aneurysm, although it was truly a multi-chamber injury involving the ascending aorta. The RCC was also injured and repaired with a pericardial patch.

There is a limited number of reports in the literature showing similar pathologies.^[5,6] Such an injury may be fatal at the time of injury, and it is quite surprising that our patient survived. In addition, he did not remember the type of knife used, although we suspected a skewer-like thin and long tool.

In conclusion, penetrating cardiac injuries may represent as unique pathologies and require major interventions. If possible, repair should be preferred, if valvular pathologies exist.

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REFERENCES

1. Connelly TM, Kolcow W, Veerasingam D, DaCosta M. A severe penetrating cardiac injury in the absence of cardiac tamponade. *Interact Cardiovasc Thorac Surg* 2017;24:286-87.
2. Morse BC, Mina MJ, Carr JS, Jhunjhunwala R, Dente CJ, Zink JU, et al. Penetrating cardiac injuries: A 36-year perspective at an urban, Level I trauma center. *J Trauma Acute Care Surg* 2016;81:623-31.
3. Abralov K, Alimov A. Short-term results of sinus of valsalva aneurysm repair. *World J Pediatr Congenit Heart Surg* 2017;8:13-7.
4. Yadav A, Mathur R, Devgarha S, Abraham V, Sisoida A. Surgery for ruptured sinus of Valsalva aneurysm: Five-year experience with 19 patients. *Turk Gogus Kalp Dama* 2014;22:729-33.
5. Esfahanizadeh J, Abbasi Tashnizi M, Moeinipour AA, Sepehri Shamloo A. undetected aorto-rv fistula with aortic valve injury and delayed cardiac tamponade following a chest stab wound: a case report. *Trauma Mon* 2013;18:95-7.
6. Kaya A, Dekkers P, Loforte A, Jaarsma W, Morshuis WJ. Traumatic aorto-right ventricular fistula with aortic insufficiency. *Ann Thorac Surg* 2005;80:2362-4.

A giant sinus node artery fistulizing to superior vena cava presenting with steal phenomenon: An unusual case report

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ABSTRACT

Congenital coronary fistulas are rare anomalies. They can be either symptomatic or asymptomatic. Management strategy for coronary fistulas differs depending on the blood flow and symptoms. Nevertheless, it should be considered a serious health problem which may result in unpleasant complications such as congestive heart failure through left-to-right shunt, myocardial ischemia through steal phenomenon, endocarditis, and aneurysmal ruptures. Herein, we present the surgical management of a young female who was referred to our clinic with exercise-induced substernal chest pain and numbness in the left upper extremity caused by a giant sinus node artery fistulizing to the superior vena cava.

Keywords: Coronary artery disease, coronary artery fistula, sinus node artery, vena cava superior.

Congenital coronary fistulas are rare anomalies with an incidence of 0.1 to 0.2%.^[1] First described by Krause in 1865,^[1] they occur as a result of anomalies in the embryology of the coronary circulation. The first surgical procedure was performed by Bjork and Crawford in 1974.^[2] The right coronary artery is the most common site of origin, and the right ventricle and atrium are the most common areas of opening.^[2] Herein, we present the surgical treatment of a rare and giant coronary fistula fistulizing to the superior vena cava through the sinus node artery.

CASE REPORT

A 31-year-old female patient was referred to our clinic with exercise-induced substernal chest pain and numbness in the left upper extremity. Her medical history revealed several treatments applied to relieve extracardiac causes-related complaints last year. Physical examination showed no pathology. Chest X-ray and electrocardiography demonstrated cardiomegaly findings. After cardiac catheterization, an extraordinary tortuous arterial structure extending from the right coronary artery to the superior vena cava was observed (Figures 1 and 2). At the preoperative examination, the aneurysmatic right coronary artery was found to give a sinus node artery branch approximately 2 cm after the separation from the

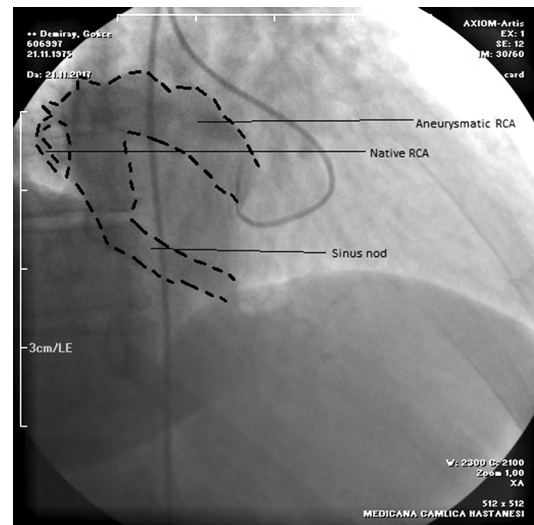


Figure 1. Appearance of fistula tract during coronary angiography.

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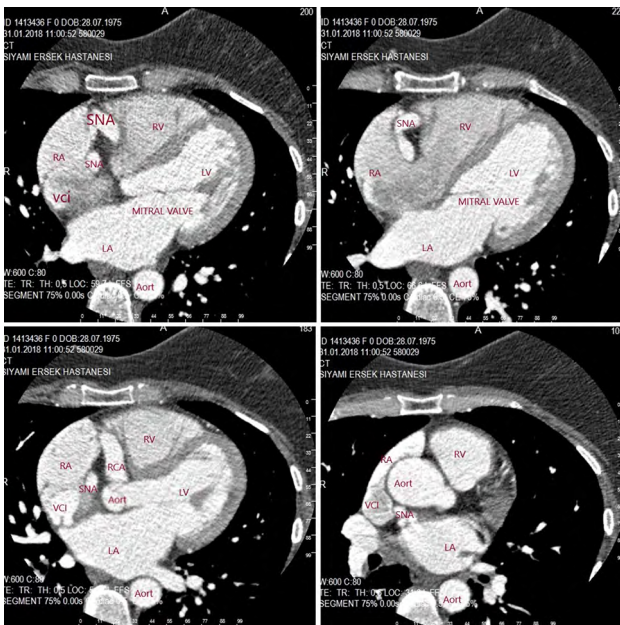


Figure 2. Computed tomography angiography showing aneurysmatic right coronary artery and sinus node artery. Right coronary artery is seen to run in normal diameter after giving the sinus node artery branch. RV: Right ventricle; RA: Right atrium; LV: Left ventricle; LA: Left atrium; SNA: Sinus node artery; VCI: Vena cava inferior.

aorta, and the diameter decreased to 4 mm there. The diameter of the sinus node artery was 1.5 cm. After running about 5 cm, it opened medially into



Figure 4. A view of resected aneurysmatic sinus node artery (lower) and an aneurysmatic segment of right coronary artery (upper).

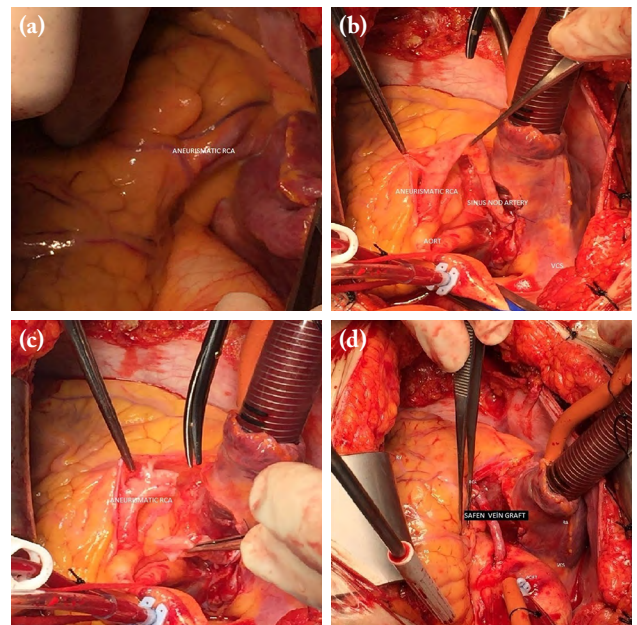


Figure 3. (a) View of the aneurysmatic right coronary artery in the anterolateral segment of the proximal aorta. (b) Aneurysmatic dilatation of the sinus node artery after exploration. (c) The appearance of the undeveloped right coronary artery after separation of the aneurysmatic sinus node artery. (d) Appearance of aorta-right coronary artery bypass with saphenous graft.

the superior vena cava, leading to a murmur of 4/6. A 2×2-cm venous aneurysm formation was observed at the opening (Figure 3a-c). Surgery was decided. A written informed consent was obtained from the patient.

Surgery was performed using standard surgical procedures with median sternotomy and bicaval cannulation. Considering the increased likelihood of rupture, the aneurysmatic proximal 2-cm segment of the right coronary artery was removed (Figures 3d and 4). The saphenous vein graft was interposed between the aorta and distal-free end of the right coronary artery (Figure 3d). The sinus node artery was also aneurysmatic and tortuous. It was excised completely and the point at which it opened into the superior vena cava was sutured. The operation was performed under cardiopulmonary bypass to avoid rhythm disturbances. After giving the sinus node artery, the right coronary artery continued about 10 cm in its normal diameter (Video 1). The postoperative course was uneventful and the patient was discharged on the postoperative fifth day.

DISCUSSION

In the presence of coronary artery disease, angina is a common finding in elderly patients. However, in younger patients and, particularly in females, angina should suggest congenital arteriovenous fistulas. Although coronary fistulas are mostly congenital, they may occur after blunt external thoracic trauma, myocardial infarction, angioplasty or cardiac surgery.^[3] Other congenital anomalies may be also accompanied by coronary fistulas with an incidence of 40%.^[3] They become symptomatic after the third decade of life. The angina is caused by coronary steal. Fistulas opening into the right heart chambers create shunts from left-to-right which may result in congestive heart failure in proportion to the size of the shunt. Such symptoms as angina pectoris, fatigue, shortness of breath, palpitation, and findings suggestive of rupture of an aneurysm, embolism, endocarditis, congestive heart failure can be seen. The degree of symptoms is proportional to the amount of physiological coronary stealing phenomenon developed by the fistula tract.^[4] The frequency of the continuous murmur, the most frequent physical examination finding, ranges from 20 to 80%.^[5] Almost half of the patients have non-specific electrocardiographic changes. Cardiomegaly can be observed on telecardiography due to an increased cardiac output.^[1] For a definitive diagnosis, selective coronary angiography is required. Thus, the origin and termination of the fistula, the path it follows anatomically, and the affected structures can be precisely visualized.

Coronary fistulas mainly originate from the right coronary artery, less frequently from the left coronary artery, or both. They were found to originate from the right coronary artery with a frequency of 19.7% in the Albeyoglu et al.,^[6] 50% in the Levin et al.,^[7] 65% in Lowe,^[8] and 51% in the Wilde and Watt^[9] series. Drainage occurs to the low-pressure heart chambers. Frequency in a decreasing order is as the right ventricle (39%), right atrium (33%), pulmonary artery (20%), left atrium, coronary sinus, vena cava superior, and bronchial arteries.^[1,7] Drainage to the left heart chambers is relatively rare (2%).^[1] Pathophysiological, myocardial ischemia or infarction occurs, when the blood to be directed to the left ventricle is directed to the low-resistance heart chambers through the fistula, creating a steal phenomenon. In fistulas which open to the left ventricle, the blood flow is frequently diastolic. Ischemia occurs, when the pulse pressure

providing the coronary flow is lowered by steal phenomenon.^[6] One of the potential complications is premature atherosclerosis, resulting in an intimal damage caused by high-volume blood flow.^[3] Our case had primarily angina symptoms and tachycardia episodes.

Nonetheless, treatment of asymptomatic fistulas is still controversial. However, most surgeons agree that they should be closed in the presence of a significant shunt or aneurysmal dilatation.^[1] Symptomatic patients need to be treated. Cardiopulmonary bypass may be required or not.^[9] Fistulas which are intramural, short, close to the sinus Valsalva and characterized by an aneurysm are often closed using cardiopulmonary bypass, while extramural and anatomically accessible ones are closed using simple ligation and resection. In addition, closure with the use of percutaneous transcatheter closure devices has become increasingly widespread, particularly in pediatric patients.^[10] Some authors do not consider surgery in asymptomatic patients, while surgery is appropriate for moderate to large-flow fistulas. Small fistulas are likely to spontaneously close as primarily or secondarily. Nevertheless, most authors consider the surgical removal of fistulas to prevent complications and sudden death risk, regardless of whether they are symptomatic or not. The reported operative mortality and morbidity rates are very low. It should be kept in mind that fistula complications may increase surgical morbidity and mortality.

In conclusion, we tried to evaluate giant sinus node artery fistula in the light of literature data. In particular, considering the causes of chest pain and arrhythmia in young patients, the consideration of coronary arteriovenous fistulas is important for the correct diagnosis.

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REFERENCES

1. Urrutia-S CO, Falaschi G, Ott DA, Cooley DA. Surgical management of 56 patients with congenital coronary artery fistulas. *Ann Thorac Surg* 1983;35:300-7.

2. Biorck G, Crafoord C. Arteriovenous aneurysm on the pulmonary artery simulating patent ductus arteriosus botalli. *Thorax* 1947;2:65-74.
3. İşcan HZ, Göl MK, Yilmazkaya B, Bardakçı B, Karagöz H, Mavitaş B, et al. Surgical treatment of congenital coronary artery fistula *Turk Gogus Kalp Dama* 2004;12:161-3.
4. Marullo AG, Sabik JF. Right coronary artery and interatrial septal aneurysms with fistulous connection to the right atrium. *Ann Thorac Surg* 2002;73:969-70.
5. Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. *Circulation* 1979;59:849-54.
6. Albeyoglu S, Aldag M, Ciloglu U, Sargin M, Oz TK, Kutlu H, et al. Coronary Arteriovenous Fistulas in Adult Patients: Surgical Management and Outcomes. *Braz J Cardiovasc Surg* 2017;32:15-21.
7. Levin DC, Fellows KE, Abrams HL. Hemodynamically significant primary anomalies of the coronary arteries. Angiographic aspects. *Circulation* 1978;58:25-34.
8. Lowe JE, Oldham HN, Sabiston DC Surgical management of congenital coronary artery fistulas. *Ann Surg* 1981;194:373-80.
9. Wilde P, Watt I. Congenital coronary artery fistulae: six new cases with a collective review. *Clin Radiol* 1980;31:301-11.
10. Kung GC, Moore P, McElhinney DB, Teitel DF. Retrograde transcatheter coil embolization of congenital coronary artery fistulas in infants and young children. *Pediatr Cardiol* 2003;24:448-53.

Successful coronary bypass surgery in a patient with hemophilia: A case report

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ABSTRACT

Hemophilia is an X-linked congenital coagulation disorder characterized by increased tendency to bleeding. Hemophilia A is an X-linked disorder resulting from a deficiency in blood clotting factor VIII, a key component of the coagulation cascade. Perioperative management is very important in this patient population. In this report, we present a 78-year-old male case with hemophilia A who underwent coronary artery bypass grafting.

Keywords: Coronary artery bypass grafting, Factor VIII, hemophilia A.

Hemophilia is an X-linked congenital coagulation disorder characterized by increased tendency to bleeding. Hemophilia A is an X-linked disorder resulting from a deficiency in blood clotting factor VIII, a key component of the coagulation cascade. Factor VIII deficiency is present in patients with hemophilia A, while factor IX deficiency is present in patients with hemophilia B. The first one is more common than the latter type of hemophilia. The incidence of hemophilia A is 1/10.000 in the world. Recent improvements in the management of hemophilia A have increased the life expectancy in these patients and, currently, the life expectancy in the hemophilia A patients is similar to the healthy population. However, age-related cardiovascular diseases have also become more prevalent in these patients. Open heart surgery poses certain challenges in patients with hemophilia A, due to the requirement of cardiopulmonary bypass (CPB) and heparinization for extracorporeal circulation. To overcome bleeding complications in the perioperative period, it is of utmost importance to identify preoperative factor levels and inhibitor levels and to administer factor replacement therapies.^[1]

In this report, we present a male case with hemophilia A who underwent coronary artery bypass grafting (CABG).

The patient underwent coronary angiography to diagnose acute anterior myocardial infarction. Diffuse three-vessel coronary artery disease was detected and the patient was advised to have CABG. His medical history revealed hemophilia A for six years, although there was no significant bleeding history during childhood and young adulthood. He underwent a right inguinal hernia repair operation six years ago. The patient was consulted to the hematology department due to a recurrent incisional hematoma. Factor VIII level was 11%. The patient was diagnosed with hemophilia A and Factor VIII replacement was administered before discharge. He had also comorbidities including chronic obstructive pulmonary disease and hypothyroidism and he was using oral levothyroxine sodium 25 µg once a day. The ejection fraction of the patient was 55%. The hematocrit level was 38%. The platelet count was 219,000/µL (reference range: 150,000-450,000/µL). The international normalized ratio was 0.99 (0.9-1.2). Factor VIII level was 20% (50-150). Factor VIII inhibitor was negative. The other biochemical laboratory test results were normal. The patient was

CASE REPORT

A 78-year-old male patient with hemophilia A was referred to the emergency department with

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consulted to the hematology department. We did not perform thromboelastography, as our consultant hematologist did not order. One hour before the operation the patient received 3,000 IU Factor VIII intravenously. Factor VIII level reached 105%. A written informed consent was obtained from the patient.

Median sternotomy was performed under general anesthesia. Intravenous heparin injection was done (300 IU/kg) after the harvesting of bypass grafts (left internal mammary artery and saphenous vein). The patient underwent four-vessel CABG. The highest activated clotting time (ACT) level was 854 sec. The temperature under CPB was 31°C. Cross-clamp time was 53 min, while CPB time was 100 min. After CPB was terminated, protamine was administered to neutralize heparin. We used tranexamic acid at a dose of 10 mg/kg bolus, 1 mg/kg/h infusion over eight h. No cell-saver was used. The patient was administered one unit of fresh frozen plasma (FFP), while no platelet suspension was given. The operation was completed in a routine fashion.

The patient had a total of 560 mL drainage during perioperatively. He stayed in the intensive care unit for three days and underwent 1,500 IU Factor VIII replacement b.i.d. for the first four days postoperatively. Over the next six days, 1,000 IU Factor VIII replacement b.i.d. was performed. Factor VIII level was 47% on postoperative Day 10. The patient was administered three units of blood transfusion and oral acetylsalicylic acid 150 mg daily as of the postoperative first day. At the time of discharge, the hematocrit level was 32%. There was no pleural or pericardial effusion or bleeding at any part of the body. He was discharged on postoperative Day 11 uneventfully.

DISCUSSION

Recent improvements in the management of hemophilia A have increased the life expectancy in these patients. The incidence of coronary artery disease has been increasing with increasing age in these patients as well as in the normal population.^[1] Interventional or surgical methods which can be applied carry a high risk of bleeding, as these patients are dependent on anticoagulant or antiaggregant therapies.

Throughout their professional lives, cardiovascular

surgeons operate a very small number of patients with hemophilia A. Therefore, both surgeons and anesthesiologists may not have adequate experience on perioperative management of these patients. The World Hemophilia Federation recommends that, in hemophilia A patients undergoing a major surgery, the preoperative target level for Factor VIII should be 80 to 100% and these levels should be maintained in the first couple days after surgery.^[2] Of note, higher levels of Factor VIII may increase the risk of thrombosis in CABG and prosthetic valves. On the other hand, cardiovascular surgeons may experience difficulties in providing hemostasis due to major alterations in the hemostatic system caused by median sternotomy, full dose heparinization, CPB, hypothermia, postoperative thromboprophylaxis, and antiplatelet or anticoagulant drugs used.

Our case did not have any bleeding complications in the perioperative period. He was administered three units of blood and one unit of FFP. To reduce the risk of graft thrombosis, the Factor VIII dose administered was gradually reduced during the postoperative period. Since the coronary anatomy of the patient was not appropriate, off-pump surgery was not chosen. However, attempts to reduce blood trauma by applying moderate hypothermia and minimizing CPB equipment were made.

During the perioperative period of patients with hemophilia A, a team of surgeons, anesthesiologists, and hematologists should follow a multidisciplinary approach.^[3] The most important point to reduce bleeding is to ensure adequate preoperative preparation. In addition, the followings are the points to be addressed: avoiding hypothermia, performing off-pump surgery, if possible, keeping CPB time short, minimizing the use of CPB equipment, and avoiding strong aspiration to reduce the trauma of shaped components of the blood. In addition, in these patients, all necessary examinations regarding the coagulation system should be done preoperatively. The opinion of the hematologist must be taken in this context. It should also be remembered to the hematologist that open heart surgeries differ from other operations due to their possible adverse effects on the coagulation system. Blood products and clotting factors which can be used in the perioperative period should be thoroughly prepared. If these preparations are not made in accordance with a multidisciplinary approach, it may be impossible to manage bleeding complications.

In conclusion open heart operations of patients with hemophilia A have significant differences and risks from other operations. Even the most experienced heart surgeons and anesthesiologists encounter these patients very rarely in their professional lives. The multidisciplinary case management that will be performed according to the recommendations of the hematologist will reduce the bleeding complications to a minimum. It should be kept in mind that excessive replacement of the Factor VIII to reduce bleeding complications may increase the risk of bypass grafts and prosthetic valve thrombosis.

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REFERENCES

1. Barillari G, Pasca S, Erice F, Livi U. Successful double bypass in a patient with severe hemophilia A: a case report. *J Thromb Thrombolysis* 2012;33:193-6.
2. Kanellopoulou T, Nomikou E. Replacement therapy for coronary artery bypass surgery in patients with hemophilia A and B. *J Card Surg* 2018;33:76-82.
3. Boğa M, Yağdı T, Dişçigil B, Büket S. Coronary artery bypass grafting in a patient with Hemophilia B (Case report). *Turk Gogus Kalp Dama* 1999;7:414-6.

Aberrant right subclavian artery aneurysm: A rare entity

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Aberrant right subclavian artery (ARSA) is the most frequent abnormality of the arch which accounts for 1% of the population.^[1] In this abnormality, the right subclavian artery leaves the left part of the aortic arch as the final branch and progresses into the right axillary region through the posterior aspect of the esophagus (i.e., from left to the right). It often progresses between the esophagus and trachea or by the anterior aspect of the trachea.^[2] This pathology is usually asymptomatic; however, it may lead to respiratory symptoms in children and difficulty in swallowing or a chronic cough in adults. In case of pressure on the esophagus, dysphagia lusoria may be observed. In case of an aneurysmatic widening of the aberrant subclavian artery in a segment close to the aorta, it is referred to as the Kommerell's diverticulum. This diverticulum may cause pressure on the tracheoesophageal region, leading to dissection/rupture due to excessive widening.^[3,4] Herein, we present an 80-year-old male patient with an ARSA aneurysm.

An 80-year-old male patient was admitted to our clinic outpatient with dizziness and fatigue which increased gradually over the past year. He also suffered from hypertension. On contrasted computed tomography scan of the thorax, ARSA was observed with fusiform aneurysmatic dilatation. A mural thrombus measured as 18 mm in the thickest part of the aneurysm wall was detected. The diameter of the aneurysmatic segment was measured as 40 mm with the thrombus and 22 mm with the patent lumen. Diffuse enlargement (fusiform aneurysmatic dilatation) in the ascending aorta diameter (42 mm) was found. In addition, intense atherosclerotic calcification was observed in the descending aorta with the aortic arch with a significant tortuosity in the descending aorta. The fusiform aneurysm was measured as 4.8 cm in

diameter at the largest site of the descending aorta. Plaque formations in the aneurysm wall were seen. The thickness of the thickest part of the plates was measured as 17 mm (Figure 1a-d). The patient was followed with medical treatment.

Antegrade cerebral protection via the subclavian or axillary cannulation is preferred during aortic arch surgeries. However, the placement of the aortic cross-clamp on the proximal aspect of the left subclavian artery may lead to serious cerebral complications in patients with an ARSA pathology. In such cases, cerebral protection can be achieved by antegrade cerebral perfusion through bilateral common carotid arteries. In patients with ARSA abnormalities and in the presence of a gastrointestinal bleeding due to delayed nasogastric or endotracheal intubation, tracheoarterial fistulas (between the trachea and ARSA) should be suspected.^[3,5]

The failure rate of transradial coronary angiography is 40% in patients with ARSA. The direction of the catheter toward the ascending aorta or to the aortic root may be difficult via the right transradial approach. The ARSA abnormality can be also observed with the absence of the right recurrent laryngeal nerve in certain cases. This is important in patients undergoing thyroid surgery. The right recurrent laryngeal nerve is absent in its normal place in the inferior pole of the thyroid gland. It is placed in the lateral aspect of the gland or in an aberrant location, and nerve

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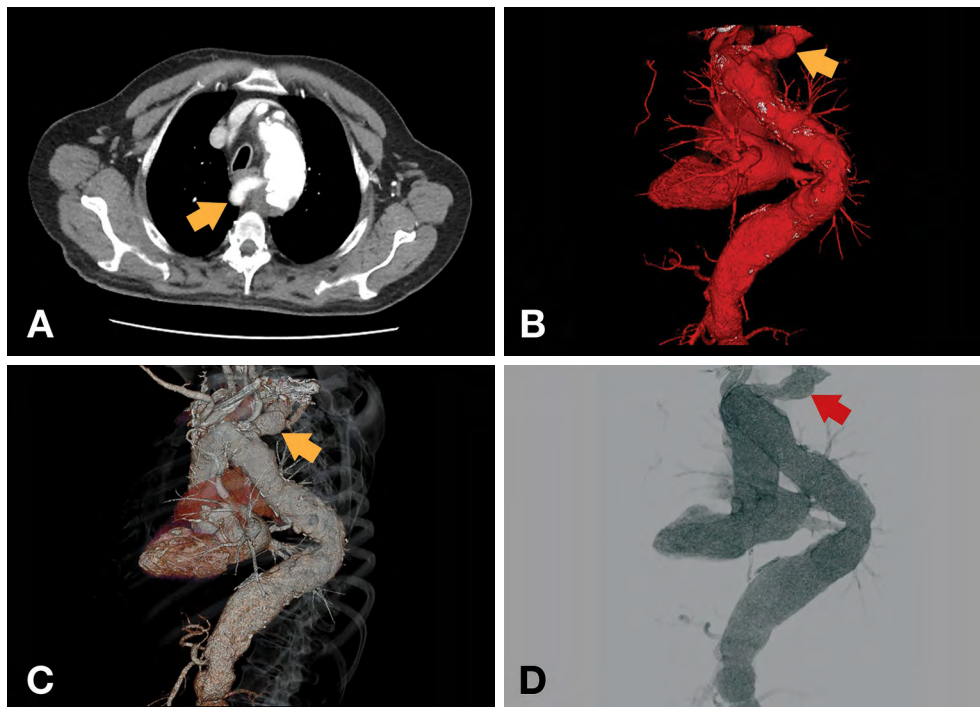


Figure 1. Thoracic contrasted computed tomography images of aberrant right subclavian artery aneurysm.

injury may occur during thyroidectomy.^[5] Again, during interventions of anterior cervicothoracic region pathologies (i.e., tumor or disc hernia) and during right thoracic outlet syndrome surgery, identifying ARSA prior to surgery would avoid vascular injuries and related bleedings.

In conclusion, previous identification of aberrant right subclavian artery is important in avoiding vascular injuries and cerebral complications in patients undergoing endovascular intervention on the aorta, aortic arch surgery, thyroidectomy, or cervicothoracic surgery.

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REFERENCES

1. Karacan A, Türkvatan A, Karacan K. Anatomical variations of aortic arch branching: evaluation with computed tomographic angiography. *Cardiol Young* 2014;24:485-93.
2. Kau T, Sinzig M, Gasser J, Lesnik G, Rabitsch E, Celedin S, et al. Aortic development and anomalies. *Semin Intervent Radiol* 2007;24:141-52.
3. Alur İ, Alihanoglu Yİ, Güneş T, Çıtışlı V. An assessment of the clinical significance of aortic arc variations. *Turk Gogus Kalp Dama* 2015;23:804-5.
4. Sierra-Galan LM, Shveid-Gerson D, Gomez-Garza G, Rey-Rodriguez A. Double incomplete aortic arch and Kommerell's Diverticulum as a cause of chronic cough. *Arch Cardiol Mex* 2015;85:158-60.
5. Polguy M, Chrzanowski Ł, Kasprzak JD, Stefańczyk L, Topol M, Majos A. The aberrant right subclavian artery (arteria lusoria): the morphological and clinical aspects of one of the most important variations--a systematic study of 141 reports. *ScientificWorldJournal* 2014;2014:292734.