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Case Report / Приказ случаја

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Two Stage Surgical Repair of Type II Acute Aortic Dissection and Aortic Coactation in a 12-year-old Child

Хируршко збрињавање у два акта 12-то годишњег детета са дисекцијом анеуризме асцедентне аорте удружене са коарктацијом аорте

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SUMMARY

Introduction Combination of acute aortic dissection associated with aortic coarctation in pediatric population is extremely rare.

Case outline A 12-year-old boy with no trauma history was admitted for chest pain. The diagnosis of of acute aortic dissection associated with aortic coarctation was established with echocardiography and Computed tomography (CT) angiography. Emergent surgery was performed: excision of the ascending aorta aneurysm with supracoronary graft replacement and preservation of native aortic valve. Subsequently, through posterolateral left thoracotomy, he underwent end to end aortoplasty for coarctation repair.

Conclusion Two stage surgery provides favorable outcome in this rare, life threatening condition in the pediatric age group. Native aortic valve was perserved and extraanatomic bypass of aortic coarctation was avoided. Further monitoring of aortic valve is mandatory.

Keywords: ascending aorta aneurysm, pediatric, coarctation

Сажетак

Увод Комбинација акутне аортне дисекције удружене са коарктацијом аорте је веома ретка у педијатријској популацији.

Приказ болесника Дечак стар 12 година, без анамнестичких података о трауми, примљен је због болова у грудном кошу. Дијагноза акутне дисекције анеуризме асцедентне аорте и аортне коарктације је постављена ехокардиографски и потврђена компијутеризованом томографијом**.** Учињен је хитан хируршки захват у виду ресекције анеуризме и супракоронарне уградње невалвуларног кондуита уз презервацију нативне аортне валвуле. Након тога учињена је аортопластика из постеролатералне торакотомије ради збрињавања аортне коарктације.

Закључак Етапно двостепено лечење овог ретког, животно-угрожавајућег стања, обезбеђује добар исход. Сачувана је нативна аортна валвула и избегнуто екстраанатомско премошћавање аортне коарктације. Неопходно је даље праћење функције аортне валвуле.

Кључне речи: анеуризма асцедентне аорте, деца, коарктација аорте

INTRODUCTION

Rupture of dissecting aneurysm is exceptionally rare, life–threatening condition in children and young adolescents [1]. Furthermore, the combination of acute aortic dissection associated with aortic coarctation in this age group is sparsely reported. Infrequent pediatric reports are mostly related to patients with Turner syndrome and other connective tissue disorders [1]. Co-existing aortic dissection and coarctation have been addressed in various ways. There are several previous reports of two-stage repair as well as one-stage repair mainly using extra anatomic by pass from ascending to descending aorta. Hereby we present, to our knowledge, the youngest patient with this condition treated with staged procedures.

CASE REPORT

A 12-year-old boy with no previous medical history was admitted for distressing parasternal chest pain, accompanied with shortness of breath and dizziness. There was no trauma history. On clinical examination radial pulses were present, whereas femoral pulses were barely palpable. A systolic murmur 2-3/6 was noted in the precordium. Electrocardiography indicated sinus rhythm with transitory ST elevation during the episodes of chest pain. Transthoracic echocardiography

demonstrated hypertrophied left ventricle, trivial incompetence (+0,5/4) of bivelar aortic valve with anulus of 2.2 cm, and significant pericardial effusion. Mitral valve was dysplastic but competent. A posterior intimal flap was detected above the aortic valve with an aneurysmal dilatation of the ascending aorta (6 cm). Furthermore, the coarctation of the aortic isthmus was demonstrated with a gradient of 50 mm Hg. A CT scane showed identical findings with the intimal flap extending bellow the origin of the innominate artery (Figure 1).

With a diagnosis of aortic dissection type II and associated aortic coarctation emergent surgery was performed through a median sternotomy. During routine anesthetic preparation arterial pressure lines were placed in right radial and left femoral artery. Upon pericardium opening 600 ml of blood was drained, grossly dilated, dissected aorta appeared. The aortic annulus was not dilated. The aneurysmatic change was abowe the sinotubular junction and stretched to 1,5 cm below the origin of innominate artery. Single arterial cannulation was performed high in the aortic arch, just bellow the innominate artery, the right atrium was cannulated with a two- stage venous cannula. Cardiopulmonary bypass was established with systemic cooling to 28°C. Preassure in the femoral artery was sufficient throughout the procedure (over 50 mm Hg). The ascending aorta was highly croos clamped, opened and cardioplegia was infused in the coronary ostia.

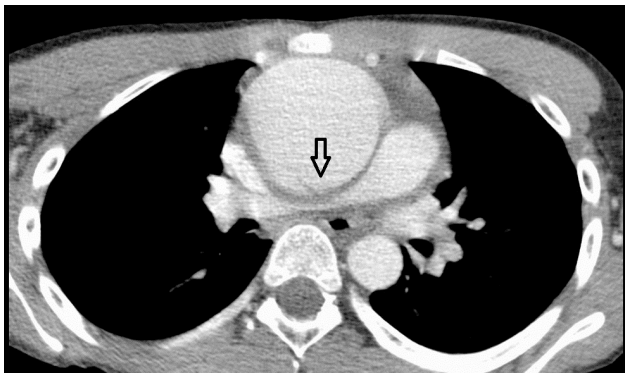

Figure 1. CT angiography: posterior intimal flap in aneurismatic ascending aorta.

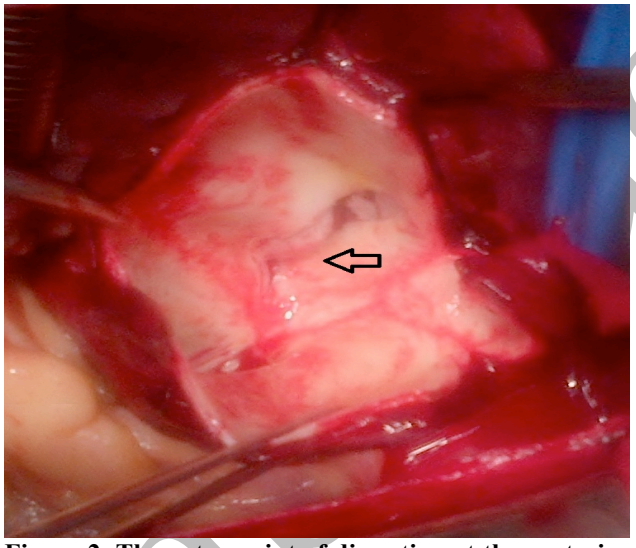

Figure 2. The entry point of dissection at the posterior aortic wall 2 cm above the right coronary ostium.

The entry point of dissection was identified at the posterior aortic wall 2 cm above the right coronary ostium (Figure 2). The aortic valve was bicuspid, though it appeared as competent and anatomically normal. After excising the aneurysm, aortic valve was preserved and resuspended with a double layer of Teflon felt inside and outside the free margin of the proximal aorta. A size 24 mm Dacron graft was anastomosed here, whereupon sealine injection into neo-aortic root demonstrated good aortic valve competence. The distal ascending aortic anastomosis was performed in the same manner with normal aortic wall just bellow the cross clamp. At the end of the procedure, after rewarming to 37°C, the patient was easily weaned off by pass.

Postoperatively he was extubated the following morning. Postoperative echocardiography demonstrated hypertrophied left ventricle with aortic valve insuffitienty +2,5/4, and mitral valve insuffitienty +1,5/4.

After achiving full recovery during the same hospitalization he was operated for aortic coarctation. Through left posterolateral thoracotomy at the fourth intercostal space aortoplasty with coarctation resection and standard "end to end" anastomosis was performed. Multiple collateral vessels of descending aorta were observed.

Posteoperativley he was extubated 5 hours after the surgery. Intensive care unit stay was 2 days. The main postoperative complication was hypertension. He was discharged ten days after the second operation on Enalapril and Presolol. Echocardiography demonstrated reduction of both mitral (+1/4) and aortic valve regurgitation (+2/4), good left ventricle function, there was no residual gradient at the place of aortoplasty.

At 24 months follow-up the patient is asymptomatic, normotensive, still on presolol. Repeated echocardiography showed no further progresion of aortic and mitral regurgitation, and no signs of left ventricle function deterioration.

DISCUSSION

The association between coarctation and aortic dissection has been described in early studies of the natural history of the aortic coarctation [2]. However reports of this condition in pediatric population are extremely rare [3]. The usual onset of dissection is the adolescent period rather than childhood, and to our knowledge, this is the youngest patient presented with this life–threatening condition. The most common predisposing factors in children are connective tissue disorders as Marfan's, Turner's and type IV Ehlers Danlos syndrome. These disorders usually have clear physical stigmata. None of those stigmata were present in the described case, our patient has been practicing water polo actively. Nevertheless, Hatzaras and colleagues reported that grueling physical activities with severe emotional stress are clear precipitating factors of acute dissection [4]. Furthermore swimming has been reported to precipitate acute aortic dissection in the absence of any predisposing factors [5]. We can speculate that mentioned factors, alongside with idiopathic dilatation and hypertension due to coarctation were the main etiological factors for dissection.

The repair of aortic dissection in the presence of coarctation comprises a few difficulties: decision on the optimal timing and sequence of the surgical repair, optimal surgical exposure, and perfusion techniques. Several surgical options have been reported. Sampath first described staged approach with initial aortoplasty followed by dissection repair [6]. On the other hand there is an opinion that dissection repair as a life saving procedure takes precedence over coarctation repair. The first single-stage repair was described by Svensson and colleagues in 1994 [7].

In the reported case we have performed two stage strategy with initial repair of dissection. Clearly, primary coarctation repair was not an option because the patient required immediate repair of

the acute dissection and relief of the cardiac tamponade. Furthermore, giving the extreme rarity of this condition, relativelly low gradient over the coarctation and unknown dissection duration time we have decided to procede with two stage strategy as a safer alternative. We have been able to maintain adequate blood flow on cardiopulmonary by pass through single arterial cannula thanks to well developed collateral vessels and relatively low gradient across coarctation. In the presence of non dilated aortic annulus, normal aortic sinuses and functional bicuspid valve we have decided not to replace the valve. Initial level of aortic and mitral regurgitation before coarctation repair raised numerous doubts about that decision. Fortunately, insufficiency of both mitral and aortic valve were reduced after the aortoplasty, and in the follow up there has been no deterioration in left ventricle function. We are aware that valve replacement will most likely prove necessary. In the meantime, the patient will hopefully complete growth and will be spared from problems related to anticoagulation therapy. One can speculate that valve–sparing aortic root implantation with a vascular graft would be better solution, but this particular operations on bicuspid valve remains challenging [8]. The mechanical valve inserted in a composite graft is known for long- term durability but these young patients are exposed to a long- term risk of thromboembolism and oral anticoagulation. One clear advantage of two stage repair is "end to end" aortoplasty which is far better solution for coarctation than extra-anatomical bypass grafting from ascending to descending aorta as a standard technique in a single stage procedures. One other option would be stenting of coarctation, nevertheless, regarding the patient's age we have chosen to give priority to surgery.

CONCLUSION

In summary, we successfully performed two stage repair with preservation of the aortic valve, replacement of ascending aorta with Dacron graft and "end to end" aortoplasty for acute type II aortic dissection with coarctation. Further monitoring is mandatory for assessing the fate of native aortic valve.

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REFERENCES

1. Fikar CR, Fikar R. Aortic dissection in childhood and adolescence: an analysis of occurrence over a 10-year interval in New York State. Clin Cardiol. 2009; 32: E23–E26
2. Abbott ME. Coarctation of the aorta of the adult type (II. A statistical study and historical retrospect of 200 recorded cases with autopsy, of stenosis or obliteration of the descending arch in subjects above the age of two years). Am Heart J. 1928; 3: 574–
3. Paparella D, Schena S, Schinosa LLT, Vitale N. One step surgical repair of type 2 acute aortic dissection and aortic coarctation. Eur J Cardiothorac Surg 1999; 16: 584–6
4. Hatzaras IS, Bible JE, Koullias GJ, Tranquilli M, Singh M, Elefteriades JA. Role of exertion or emotion as inciting events for acute aortic dissection. Am J Cardiol. 2007; 100: 1470–1472

5. Edwin F, Aniteye EA, Sereboe L, Frimpong-Boateng K. Acute aortic dissection in the young: distinguishing precipitating from predisposing factors. Interact Cardiovasc Thorac Surg. 2009; 9: 368.
6. Sampath R, O'Connor WN, Noonan JA, Todd EP. Management of ascending aortic aneurysm complicating coarctation of the aorta. Ann Thorac Surg 1982; 34: 125–31.
7. Svensson LG. Management of acute aortic dissection associted with coarctation by a single operation. Ann Thorac Surg 1994; 58: 241–3
8. Kallenbach K, Karck M, Pak D, Salcher R, Khaladj N, Leyh R, et al. Decade of aortic valve sparing reimplantation: are we pushing limits too far? Circulation 2005; 112: I253–9.

