A Giant Sternum-eroding Aneurysm of the Ascending Aorta and Aortic Arch

Gigantska anevrizma navzgornje aorte in aortnega loka z erozijo prsnice

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Ključne besede:
anevrizma aorte, insuficienca aortne zaklopke, zunajtelesni obtok, cirkulatorni zastoj, hipotermija

Key words:
aortic aneurysm, aortic valve insufficiency, cardiopulmonary bypass, circulatory arrest, hypothermia

Citirajte kot/Cite as:
Zdrav Vestn 2011; 80: 593–98

Abstract

Background: Giant aneurysms of the ascending aorta, defined as aneurysms of more than 10 cm in diameter, are a rare finding. They represent a high risk of dissection or rupture and can also compress the surrounding structures and organs. Generally, the only effective treatment is surgery.

Case report: In this report we present a case of a giant sternum-eroding aneurysm of the ascending aorta and aortic arch in a progressively dyspnoic 34-year old female and describe a stepwise surgical approach as the optimal treatment.

Conclusion: Surgical treatment of giant aneurysms of the ascending aorta carries high morbidity and mortality particularly when compressing the surrounding structures or causing bone erosion. A stepwise surgical approach with the establishment of CPB and hypothermia prior to sternotomy, precise surgical technique, and meticulous postoperative care are the factors which significantly improve the safety and efficacy of the procedure and all contribute to a better outcome.

Izvleček

Uvod: Anevrizme navzgornje aorte, ki v premeru presegajo 10 cm, imenujemo gigantske anevrizme in so zelo redke. Spremlja jih velika verjetnost razpoke ali disekcije, nevarne pa so tudi zato, ker lahko zaradi svoje velikosti pritiskajo na sosednje strukture in organe ter jih poškodujejo. Zdravljenje je v večini primerov kirurško.

Prikaz primera: V prispevku predstavljamo primer uspešne operacije gigantske anevrizme navzgornje aorte, ki je pri 34-letni bolnici povzročala postopno napredovanje občutka težke sape in zaradi svoje velikosti povzročila erozijo in stanjšanje prsnice.

Zaključek: Kirurško zdravljenje gigantskih anevrizem navzgornje aorte spremlja visoko tveganje, še posebej kadar pritiskajo na sosednje strukture. Stopenjski kirurški pristop z vzpostavitvijo zunajtelesnega obtoka in hipotermije še pred sternotomijo, natančna in primerno hitra kirurška tehnika ter dobra pooperativna oskrba enakovredno pripomorejo k varnosti in učinkovitosti zdravljenja ter odločilno prispevajo k boljšim rezultatom.

Introduction

An aortic aneurysm is a localized dilatation of the aorta to more than 1.5 times compared to the normal aortic diameter at the same anatomical level.[1] Aneurysms of the thoracic aorta consistently increase in size and progress to serious complications including rupture, which is usually fatal. A giant aneurysm of the ascending aorta, defined as an aneurysm of more than 10 cm in diameter, is a rare finding. Besides representing a threat of dissection or rupture,

Prispelo: 29. okt. 2010,
Sprejeto: 8. apr. 2011

they can also compress the surrounding structures or cause sternal erosion.[2,3]

Aortic aneurysms are the 13th leading cause of mortality in the United States.[4] The incidence of thoracic aortic aneurysms is estimated to be 5.9 cases per 100,000 person-years.[5]

In this report we present a case of a giant sternum-eroding aneurysm in an otherwise healthy 34-year old female.

Case Report

A 34-year-old woman was referred to our institution due to gradually developing dyspnea.

An angio-CT scan showed a giant aneurysm involving the ascending aorta and the proximal part of the aortic arch with the greatest diameter of 14 cm. The aneurysm completely occupied the retro-sternal space, causing sternal erosion in its full length. No sign of dissection was present. Preoperative echocardiography revealed a bicuspid aortic valve with a concomitant severe regurgitation and preserved left ventricular function.

After the informed consent was obtained, the patient was taken to the cardiovascular operating room and anesthetized in the usual fashion.

After heparin at a dose of 3 mg/kg was administered, the right axillary artery and right femoral vein were cannulated and the cardiopulmonary bypass (CPB) initiated. The patient was cooled to 23°C before sternotomy in order to avoid the patient's exsanguination in case of direct entry into the aneurysm during sternotomy and the resultant immediate need for circulatory arrest.

When the desired temperature was achieved and the aneurysm decompressed by means of CPB, the division of the sternum was performed with an oscillating saw without entering the aneurysm. A markedly thinner sternum due to bone erosion was found. The aneurysm involved the ascending aorta and the aortic arch. The CPB was discontinued, the aneurysmal sac opened and a bicuspid aortic valve encountered. Myocardial protection was achieved by intermittent retrograde cold blood cardioplegia. The aortic arch was replaced with a tri-

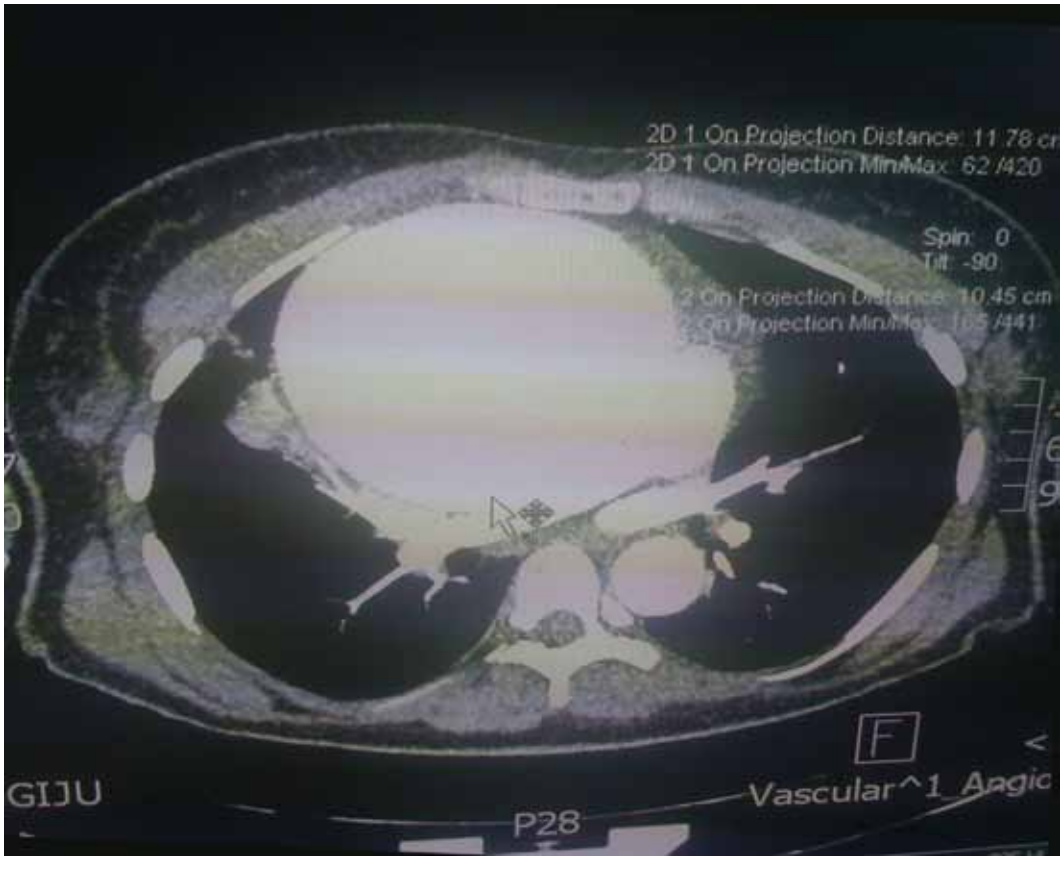

Figure 1 and 2: Preoperative angio-CT showing a huge aneurysm larger than the heart size

furcated Dacron prosthesis. Distal tubular and all branch anastomoses were constructed under circulatory arrest for a total of 22 minutes at 23°C with the head packed in ice. After the anastomoses were completed, the proximal part of the trifurcated graft was clamped, CPB circulation resumed, and the patient rewarmed to 32°C. Because of a very short duration of the circulatory arrest (22 minutes) we did not perform any selective brain perfusion.

Next, attention was focused on the aortic valve. It was a bicuspid valve with aneurysmatic enlargement of the sinuses of Valsalva and the aortic annulus. Because the probability of a succesful valve repair with a valve-sparing procedure was low, we decided to replace the valve. A Bentall procedure implanting a mechanical valved conduit (St. Jude Medical, St. Paul, Minnesota, USA) with coronary button implantation was performed. Samples of aortic valve leaflets and aneurysmal wall were sent for histology. Finally, the conduit and the trifurcated graft were anastomosed in an end-to-end fashion. After the cardiac chambers were deaired, the cardiac activity restored, and complete rewarming achieved, the patient was gradually weaned from the CPB. Heparin was reversed with protamine and the patient decannulated. When sufficient hemostasis was achieved, sternal wound was closed in the usual manner. During the surgical procedure and postoperative period the patient required a total of 6 units of blood, 12 units of fresh frozen plasma, and 5 pools of platelets. The postoperative course was uneventful. Postoperative echocardiographic evaluation documented perfect restoration of the aortic lumen. The histopathologic examination revealed thinner elastic lamellae of the aortic media with numerous foci of cystic medial degeneration. The patient was discharged on the twelfth postoperative day without any neurological deficit.

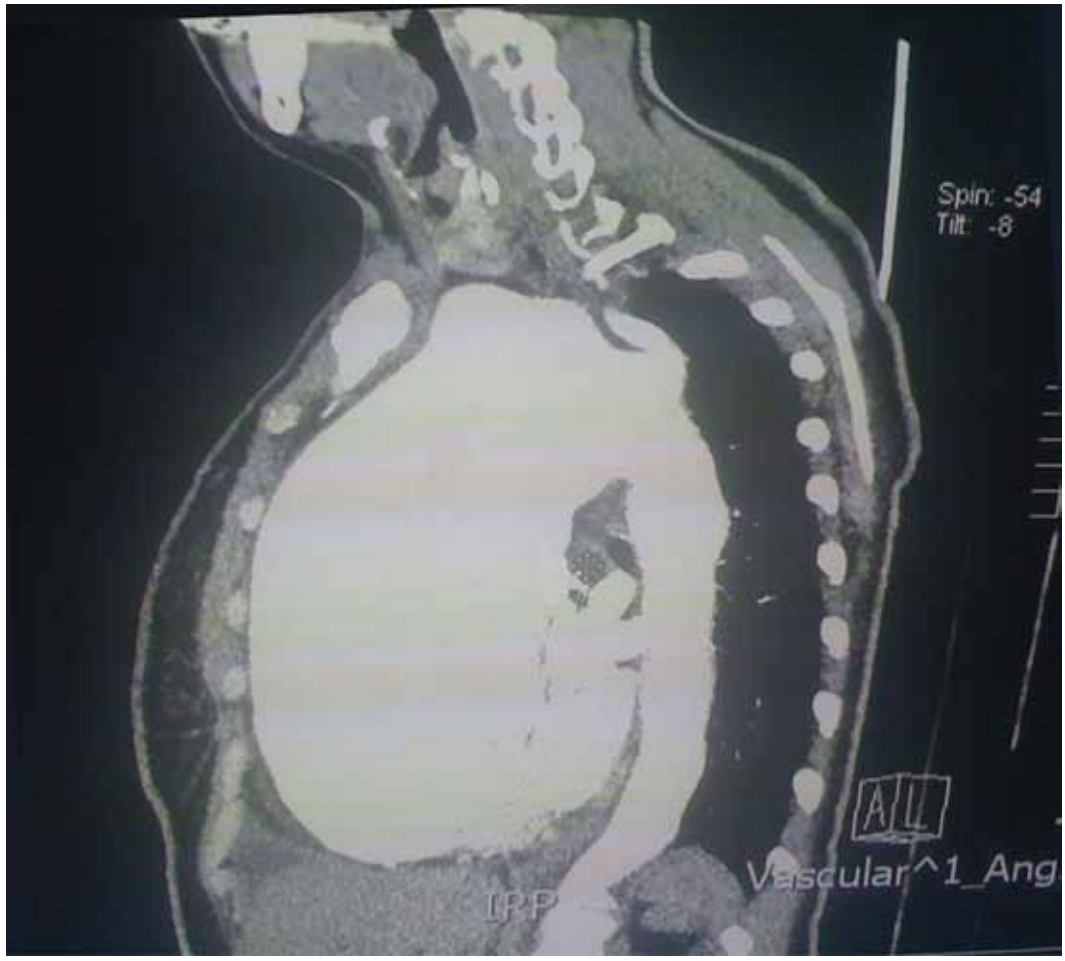

Figure 2

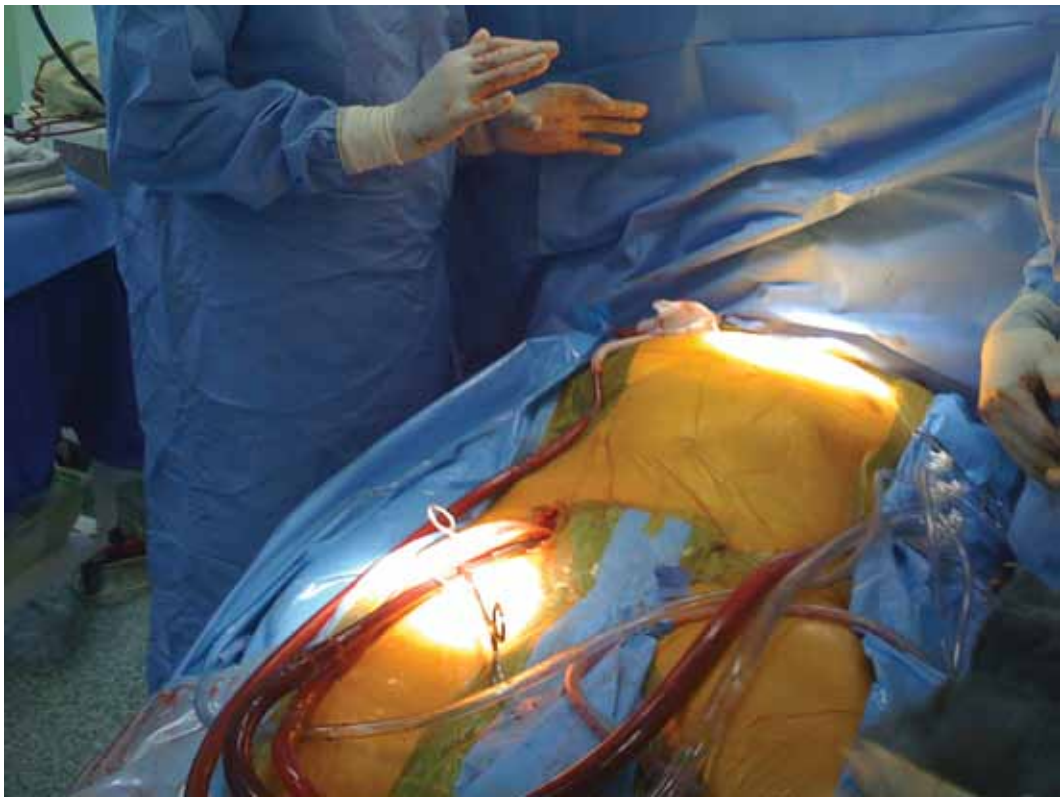

Figure 3: CPB and cooling initiated via cannulated right axillary artery and right femoral vein prior to sternotomy

Discussion

Giant ascending aortic aneurysms are rare and nowadays, as diagnostic tools improve, seen less frequently. However, when they do occur, the presentation tends to range from being asymptomatic to a dramatic medical emergency. Various etiology is well known to be associated with thoracic aneurysms. Unlike aortic aneurysms in older adults that are primarily associated with hypertension, smoking, and hypercholesterolemia, aneurysms in young individuals generally occur in the setting of an inherited disorder.[6] Among genetic disorders, Marfan's syndrome, and bicuspid aortic valve are the most prominent. The frequency of Marfan's syndrome is estimated to 1 of 5000 live births and has been associated with more than 85 mutations at one locus on the fibrillin gene on chromosome 15.[7] On the other hand, the genetic basis of the bicuspid aortic valve is largely unknown.[8,9] However, the bicuspid aortic valve represents the most common congenital heart abnormality, affecting up to 2 % of the population.[8] Less common genetic disorders for dilatation of the ascending aorta include Loeys–Dietz syndrome, Ehlers–Danlos syndrome type IV, arterial tortuosity syndrome, and autosomal dominant polycystic kidney disease.[10] Other etiologies include Giant cell, Takayasu's and Lupus arteritis and various infectious diseases (e.g. HIV associated vasculitis, tuberculosis, and syphilis).[10-12]

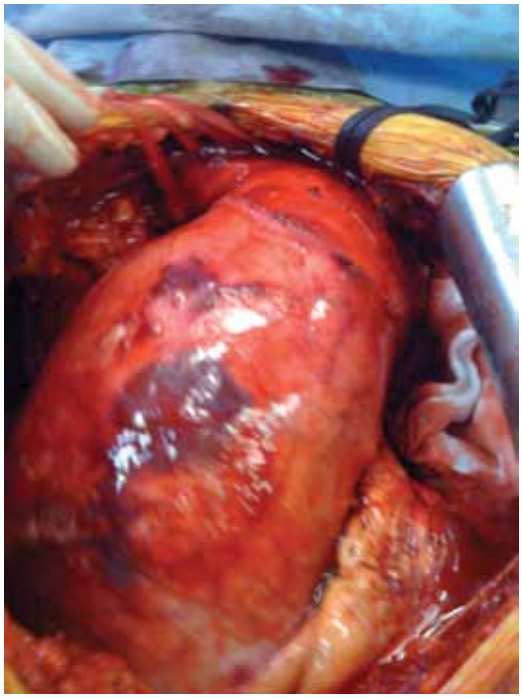

Figure 4: Intraoperative view of the aneurysm of the ascending aorta and the aortic arch

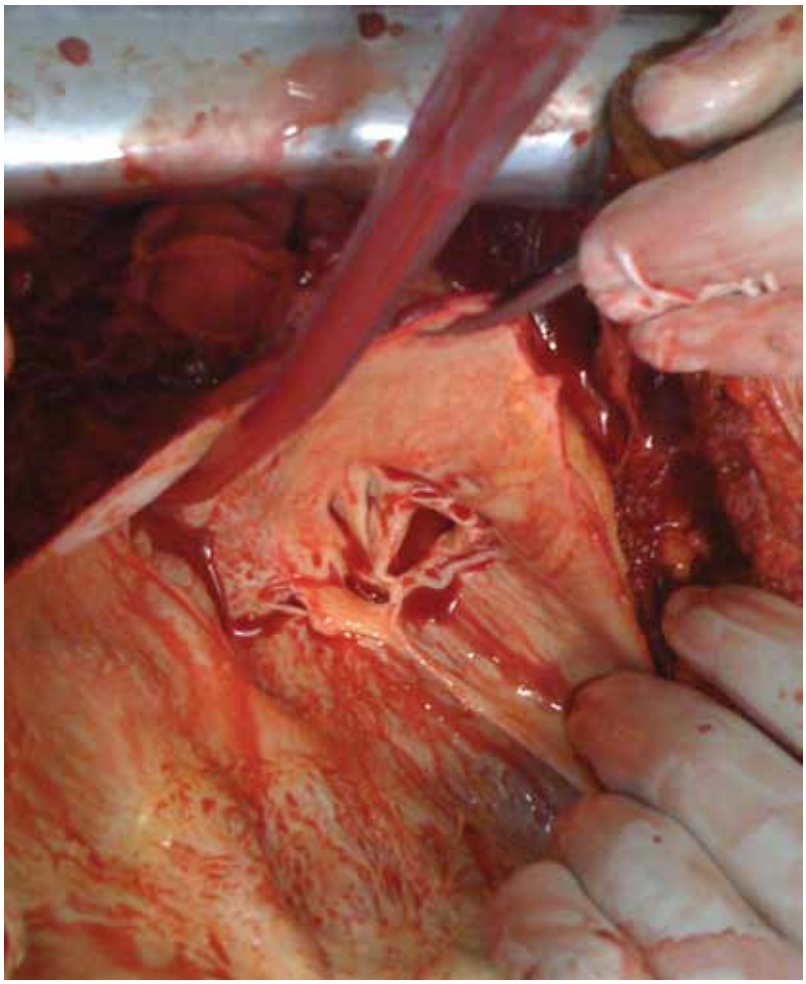

Figure 5: Opened aneurysmal sac with the view of the bicuspid aortic valve

While the symptomatic aneurysms must be resected regardless of their size, it is crucial to estimate the balance of risk versus the benefit from resection in asymptomatic cases. In the natural history described by Elefteriades,[7] the aneurysmal ascending aorta grows at an average rate of 0.07 cm per year, however, the larger the aneurysm, the faster it grows. The critical hinge point for rupture or dissection was found to be at 6 cm. By the time the aneurysm reaches this size, 31 % of patients will suffer rupture or dissection of the aorta.[7] Similarly, Coady et al.[13] reported a 4.3-fold increased risk of rupture or dissection in an aneurysm of 6.0 to 6.9 cm in diameter, compared to an aneurysm of 4.0 to 4.9 cm in diameter. They also observed a more than twofold annual growth rate (0.16 cm) in aneurysms with a diameter greater than 8 cm compared to those with the diameter of less than 4 cm. According to the recent guidelines from the American College of Cardiology/American Heart Association[14] in patients with bicuspid aortic valve, surgery is indicated when the aneurysm reaches 5 cm or 4.5 cm if concomitant aortic valve regurgitation or stenosis is present.

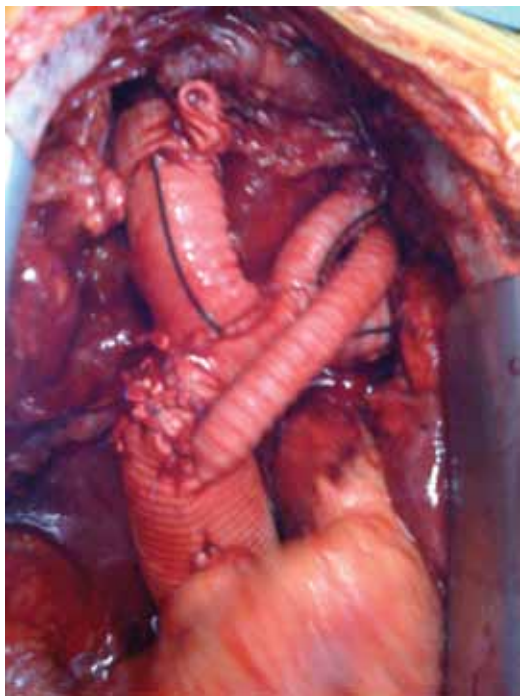

Figure 6: Final reconstruction

The surgical treatment of these patients is technically challenging and carries a high morbidity and mortality risk, especially when compression or erosion of the surrounding structures is present. Bone erosion and costal osteolysis caused by aneurysms are attributed to an enhanced activity of osteoclasts due to irritation by the pulsating mass.[15] In their most severe form, they present as pulsatile swelling of the chest wall, which may even rupture externally.[15] However, most commonly, bone erosion is limited to thinning the sternum to a varying degree. This was also true of our patient, since the sternum was thinned to approximately 5 mm in the most affected area.

It is difficult to lay down the principles for the treatment of giant ascending aortic aneurysms as they are rare and require treatment on an individual basis. In these high-risk patients, issues such as approach to the aneurysm, establishment of cardiopulmonary bypass, control of bleeding, and postoperative care have a major impact on the final outcome. In our opinion, a cautious and stepwise surgical strategy we have

implemented has many advantages. It allows a safe and effective surgical management of a giant ascending aortic aneurysm with a short circulatory arrest, thereby preventing potential operative complications and ensuring an expeditious postoperative recovery.

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