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Acute Type A Aortic Dissection – A Case Beyond the Guidelines

Акутна тип-А аортна дисекција – Случај изван препорука

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SUMMARY

Introduction There are not so many cases among acute type-A aortic dissection survivors deserving the epithet "incredible." Here we present such a case followed-up for more than five years.

Case outline A 48 years old male with acute type A aortic dissection, complicated with cardiac tamponade and severe aortic valve regurgitation, was submitted to emergent surgical treatment. Distal reconstruction was performed by complete aortic arch replacement with "elephant trunk" extension and separate arch branch bypasses, while the proximal reconstruction was done with Bentall procedure. Total of 11 anastomoses was necessary to complete this procedure. Straight profound hypothermic (18° C) circulatory arrest, with a saturation of the venous blood from the jugular bulb of 97%, lasted 133 minutes! The patient was discharged stable without any neuro-cognitive deficit. Two years later, he was admitted with late prosthetic valve endocarditis and subvalvular abscess. Good response on treatment with efficient combined antibiotics and stabile hemodynamic allowed us to avoid barely feasible re-do surgery. Subvalvular myocardial abscesses evolved into periprosthetic pseudoaneurysms without infectious, thromboembolic, or hemodynamic deterioration. The patient is still alive and stable, more than four years after this event.

Conclusion Fortunate outcome of these life-threatening conditions is a reason to reconsider our understanding of cerebral function and metabolism during the profound hypothermic circulatory arrest, and emphasizes the importance of measuring individual patient response against disease treatment guidelines, as we did, treating the late, complicated prosthetic valve endocarditis with medicaments, instead of high-risk surgery.

Keywords: aortic dissection; circulatory arrest; brain protection; prosthetic valve endocarditis

САЖЕТАК

Увод Не постоји много пацијената који су преживели акутну тип-А дисекцију аорте, а чији исход би оправдао епитет „невероватног случаја“. Циљ нам је да прикажемо један такав случај, који смо пратили више од пет година након операције.

Приказ случаја Мушкарац стар 48 година, са акутном тип-А дисекцијом аорте, компликованом тампонадом и тешком аортном регургитацијом хоспитализован је ради неопложне кардиохируршке интервенције. Дистална реконструкција је подразумевала комплетну замену лука аорте са „elephant trunk“ екстензијом уз одвојене „bypass“ реконструкције за све три гране лука, док је проксимална реконструкција захтевала извођење Bentall операције. Укупно 11 анастомоза је било неопходно за комплетирање ове процедуре. Директан дубоки хипотермни (18° C) циркулаторни застој, са сатурацијом венске крви из југулатног булбуса од 97%, трајао је 133 минута! Пацијент је отпуштен у стабилном стању без неуро-когнитивних дефицита. Две године касније, хоспитализован је под сликом ендокардитиса вештачке валвуле са субвалвларним апсцесима. Добар одговор на терапију комбинованим ефикасним антибиотикима уз стабилну хемодинамику, омогућио нам је да избегнемо реоперацију високог ризика. Субвалвларни апсцеси су еволуирали у перипротетичну псеудоанеуризму без знакова инфекције, тромбо-емболијских и хемодинамских погоршања. Пацијент је жив, у стабилном стању, више од четири године након ове компликације.

Закључак Исход лечења овог пацијента је довољан разлог да преиспитамо наше разумевање церебралне функције и метаболизма у условима током дубоког хипотермног циркулаторног застоја, као и да нагласимо важност индивидулане процене ризика сваког болесника, чак и кад се он значајно разликује од актуелних препорука, као у овом случају, кад је компликовани ендокардитис вештачке валвуле лечен антибиотикима, уместо хируршки.

Кључне речи: аортна дисекција; циркулаторни застој; протекција мозга; ендокардитис вештачке валвуле

INTRODUCTION

Experienced surgeons would agree that each case of acute type A aortic dissection (A-AAD) is peculiar. This emergent entity is well known as a dreaded disease with many faces,

classified among the five most common misdiagnoses. [1] Surgical treatment strategies are permanently evolving to ensure appropriate reconstruction, with the lowest possible brain and visceral organ damage. Accordingly, mortality and morbidity rates, as well as long-term results, significantly improved [2, 3, 4]. Yet, there are not so many cases among survivors deserving the epithet "incredible." Here we present such a case of the A-AAD, followed-up for more than five years. The purpose of this report is not to recommend our treatment strategy, but to recall and expose all kind of concerns we had, celebrating the unexpectedly fortunate outcome of this "drama in two acts."

CASE OUTLINE

The First Act: Acute Type A Aortic Dissection

A 48 years old male (195 cm, 92 kg), with a history of uncontrolled hypertension, was admitted with ongoing chest pain, hypotension (80/40 mmHg, sinus tachycardia 110/min), spontaneously breathing, and somnolent, with A-AAD, verified by multislice detector computed tomography (MDCT), complicated with cardiac tamponade and moderate-to-severe aortic valve regurgitation. Except for hypertension, no other known risk factors for A-AAD were present. Pain-to-table time was six hours, while diagnosis-to-table time was 45 minutes.

Median sternotomy and left common femoral artery access were done simultaneously, to relieve severe tamponade and ensure fast arterial line placement. Following the massive coagulum removal from the pericardial sac, the two-stage venous cannula and left atrial vent was placed. Retrograde extracorporeal circulation was initiated, and the patient was cooled down to 18° C (straight profound hypothermia), under the alpha-stat protocol, without aortic clamping and cardioplegia. Bilateral ice-pads were placed over the carotid arteries and no additional pharmacological measures for brain protection were used. Cerebral blood

oxygenation was monitored by near-infrared spectroscopy and saturation of the venous blood from the jugular bulb before the arrest was 97%. Aortotomy revealed multiple irregular tears in the ascending aorta and the arch, destroying orifices and proximal parts of all brachiocephalic arch branches. The left subclavian artery and proximal descending aorta were nearly occluded by the false lumen. At this point, there was no chance to change our initial straight profound hypothermic circulatory arrest and „the-arch-first“ strategy, so that we had to go on with „hostile“ anastomoses at the distal aorta and arch branches. The distal aortic anastomosis was performed with inverted (26 mm) tubular Dacron graft, leaving an elephant-trunk extension of 7 cm in descending aorta. Arch reconstruction necessitated resection of proximal 3-4 cm of all three branches (all far above the left brachiocephalic vein) and interposition of small (10 and 8 mm) tubular Dacron grafts. Anterograde reperfusion was started by direct arterial cannulation of the distal aortic graft. (Figure 1A) Total straight profound hypothermic circulatory arrest (PHCA) time, without any cerebral perfusion, lasted for 133 minutes! Proximally, two of three commissures were detached so that Bentall procedure, using Valsalva Dacron composite graft (23 mm valve, 24 mm graft), was necessary. Overall, a total of 11 anastomoses were necessary to complete this procedure. Hemostasis was perfect and supported by fibrin sealant on each suture line. The patient was easily weaned from cardio-pulmonary bypass, without any haemodynamic support.

Extremely difficult pathoanatomy of the arch and extremely long PHCA time without any brain perfusion did not leave us any objective reason to be optimistic. Yet, on the fifth postoperative day, the patient woke up with no neurological or cognitive deficits. Brain MDCT was completely normal. (Figure 1B) Two weeks after the operation, significant pericardial and left pleural effusions were drained under fluoroscopic control, with no residual effusions on discharge. Control echocardiography (ECHO) revealed normal structural and functional parameters of the aortic valve and the left ventricle. He was

discharged with blocker (Presolol[®] 2x25mg), ACE-inhibitor (Zorkaptil[®] 2x6.25mg), Aspirin[®] (1x100mg), anticoagulant (Farin[®] with target INR 2-3), Ibuprofen (Brufen[®] 3x600mg), Colchicine[®] (2x0.5mg), combined diuretics every other day: Furosemide (Lasix[®] 1x40mg) plus Spironolactone (Aldactone[®] 1x25mg), Cefixime (Pancel[®] 1x400mg, one week) and Lansoprazole (Sabax 2x30mg). Except for occasional hypertensive episodes, patient's condition and laboratory findings were excellent during the next two years follow-up. He went back to his usual physical and intellectual activities and actively took part in the "Aortic Disease Awareness Day" (ADAD). [5]

The Second Act: Prosthetic Valve Endocarditis with Subvalvular Extension

Almost four years ago, after respiratory infection with prolonged fever (up to 40.2°C), fatigue, headache, and myoarthralgia, he was examined at Urgent Centre and discharged with Erythromycin[®] (4x500mg). Transthoracic ECHO did not reveal any prosthetic valvular pathology. A week after, he was admitted at the Clinic for infectious diseases, with same symptoms and positive bio-humoral syndrome (Le 18.7, SE 74, Fib 6.3, CRP 177.6). The ECHO on admission revealed the presence of prosthetic valve endocarditis (PVE) with semicircular subvalvular abscess, affecting interatrial and interventricular septum. Repeated blood cultures were positive for coagulase-negative staphylococci (CoNS) and therapy was conducted combining different efficient antibiotics (Vancomycin, Levofloxacin, Ciprofloxacin, Linezolid, Teicoplanin, Amikacin). Subsequent ECHO and MDCT examinations revealed an emptied abscess cavity in a form of the periprosthetic pseudoaneurysm (with a maximal diameter of 20 mm) and normal prosthetic valve function. (Figure 2A) Both his good response to the treatment and our reluctance for re-do surgery resulted in consent for "watchful-waiting" strategy and surgery just in case of any deterioration. After two months of therapy, he was discharged in a good clinical and

hemodynamic condition with much better laboratory parameters (Le 6.7, SE 62, Fib 5.6, CRP 10.4).

During the next three years, there was no relapse of the PVE. Control ECHO and MDCT scans were scheduled on a six months basis and revealed persistent, stationery periprosthetic pseudoaneurysm without any signs of thrombosis. The valvular function was normal as well as other functional parameters of the left ventricle. (Figure 2B) Again, the patient is physically and intellectually active, still actively supporting the ADAD on each 19th of September.

Written consent for the publication of this case report and any accompanying images was obtained from the patient.

DISCUSSION

Acute aortic dissection, critical preoperative condition, extremely difficult pathological anatomy affecting ascending aorta, arch and branches, complex surgical reconstruction (i.e. 11 anastomoses), straight PHCA far above tolerable time limits (i.e. 133 minutes), postoperative pericardial and pleural effusion – all these things ("the first act") our patient survived without any serious consequence. Moreover, two years later ("the second act"), he also survived another life-threatening condition – PVE with perivalvular extension treated only with medicamentous therapy, with stabile course three years after. Both separately and especially in combination, these two "acts" indeed deserve the epithet "incredible". So far, there are no literature reports of similar cases.

As for "the first act", we want to emphasize the importance of time in the management of patients with suspected A-AAD [6] with or without malperfusion syndrome. [7] Accordingly, with a pain-to-table time of six hours and diagnosis-to-table time of only 45 minutes, we have managed to avoid serious malperfusion and multi-organ system failure. As

for the operative strategy, it is well-known that cardiopulmonary bypass (CPB) induced PHCAs in humans, lasting >40 minutes significantly increase the incidence of neurological deficits, while those of >60 minutes significantly increase mortality caused by cerebral damage. [8-10] Swenson has reported 20.8% mortality and 14.6% stroke rates in 48 patients with PHCA 60-120 minutes. [11] We could not find any literature report of straight PHCA >120 minutes in humans. Yet, there are some reports of complete recovery after 6-8 hours of cardiac arrest from deep accidental hypothermia, but such cases could not be compared with CPB induced PHCAs. [12, 13] In addition, there were experimental studies, which did not find any cerebral pathologic changes or behavioural disorders after 90 and 120 minutes of PHCA in dogs, except hippocampal apoptosis following 120 minutes of PHCA at 15° C. Such a good ischemic tolerance may be attributed to non-thoracotomic CPB and the absence of hemodilution [14] Repeated CT scans did not reveal any brain damage in our patient. Though we did not use any tools to precisely measure his neurocognitive function, neither we nor his family members could proclaim it impaired in any aspect. [15] Subsequent critical appraisal revealed that antegrade cerebral perfusion could be established earlier, by reconstructing the brachiocephalic trunk as the first step. We missed this opportunity being distracted by an impressive arch and branches pathology.

As for “the second act,” class I, level of evidence C recommendation for surgery in PVE claims: “Surgery is indicated for patients with PVE who present with complications, for example, abscess formation”. [16] Two years after the initial surgery, our patient developed late CoNS PVE with paravalvular extension. During this hospital stay, we were consulted daily to eventually decide for the re-do surgery. Our “out-of-the-box” watchful waiting attitude did not have any support in contemporary recommendations or some other literature reports. [16, 17] Good response on combined antibiotic therapy, stable general and hemodynamic condition, on the one side, and complex underlying pathology requiring very

high-risk surgery, on the other side, were the principal reasons for such approach. Not only the standard hazards of re-do surgery raised our concerns, but also some specific circumstances, such as the presence of coronary artery buttons (sealed with fibrin glue), uncertain geometry and histology of the periprosthetic pseudoaneurysm and adjacent myocardium. Guideline based surgery recommends complete reconstruction, including exclusion/excision of the pseudoaneurysm, prosthetic aortic valve and ascending aortic graft replacement with re-implantation of coronary artery buttons (or Cabrol modification). [16] In the presence of scar tissue enhanced with fibrin sealants, such demanding procedure would be hardly feasible. A few years later, the report by Saitto and associates, was the only one to compare with, to justify our concerns and decision we made. [18] Unlike their one-year follow-up experience, our patient is still alive with satisfactory clinical condition and hemodynamics, more than three years after the PVE (13th of October, 2016) and more than five years after the initial operation (13th of October, 2014).

A single case could never be a reason to build up an attitude or recommendations based on it. This particular case is definitively a reason to reconsider our understanding of cerebral function and metabolism during the PHCA conditions. A fortunate outcome with no neurological and/or cognitive deficits after 133 minutes of PHCA deserves it. Besides, this case emphasizes the importance of measuring individual patient response against disease treatment guidelines, as we did, deciding to treat the late, complicated CoNS PVE with medicaments, instead of high-risk surgery, fulfilling the Hippocratic injunction: "Primum non-nocere (i.e. Above all, do no harm)". [19]

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Conflict of interest: None declared.

Paper accepted

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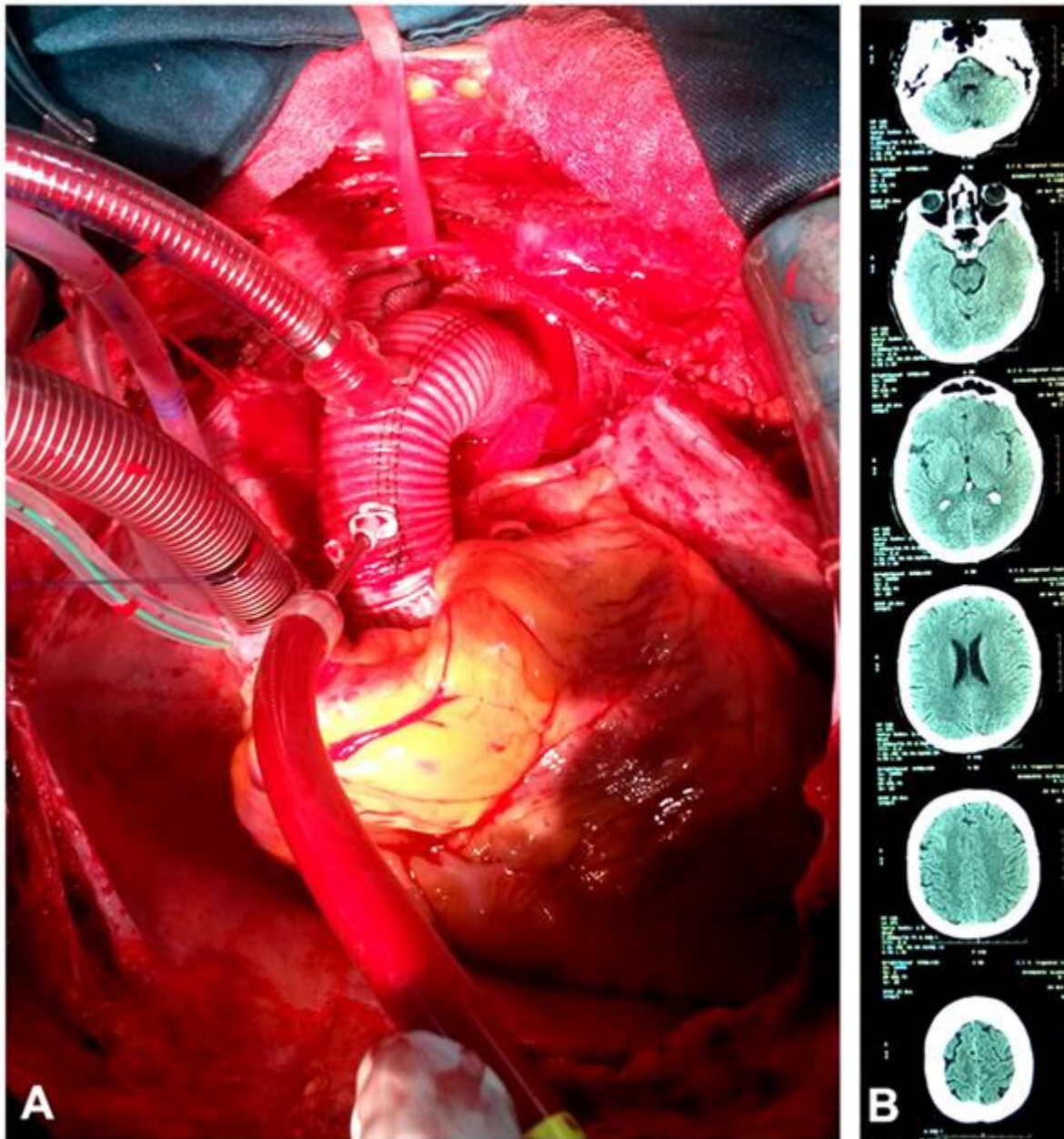


Figure 1. **A** – Completed procedure (11 anastomoses): Proximal Bentall + Complete aortic arch and distal reconstruction (separate arch branches bypasses, distal „elephant trunk“ descending aortic extension); **B** – Postoperative brain multidetector computed tomography: Absence of neurological and/or cognitive deficits. Normal multidetector computed tomography

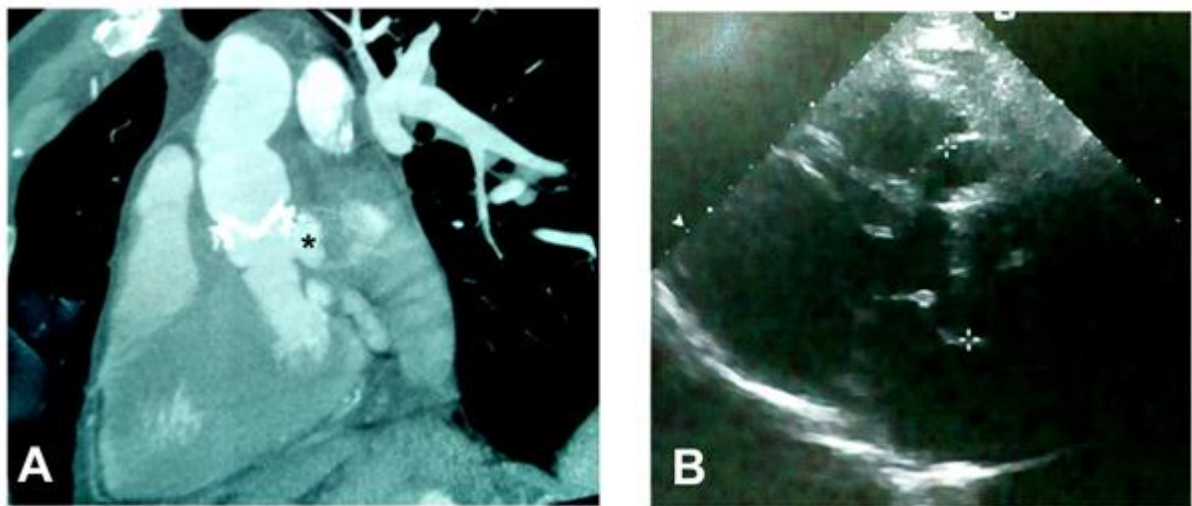


Figure 2. **A** – Postoperative chest multidetector computed tomography: An emptied subvalvular abscess (black asterisk); **B** – Postoperative ECHO: Periprosthetic pseudoaneurysm (white asterisks)