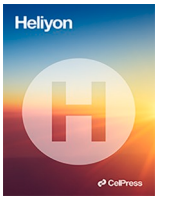
Heliyon 10 (2024) e25235

Contents lists available at ScienceDirect

Heliyon

journal homepage: www.cell.com/heliyon



Case report

Aortic arch rupture in a patient with Marfan syndrome and previous aortic root repair: A stepwise approach to intraoperative catastrophe

Talon Jones [a], Joel Price [b], Sean R. McLean [a,*]

[a] *Vancouver Acute Department of Anesthesiology, Vancouver General Hospital, JPP3 Room 3400, 899 West 12th Avenue, Vancouver, British Columbia, V5Z- 1M9, Canada*

[b] *Division of Cardiac and Aortic Surgery, VGH Gordon and Leslie Diamond Health Centre, 2775 Laurel St., 9th floor, Vancouver, British Columbia, V5Z 1M9, Canada*

A R T I C L E I N F O

Keywords:
Marfan syndrome
Aortic rupture
Massive transfusion
Resuscitation
Cardiac anesthesia

A B S T R A C T

Marfan syndrome (MS) is an autosomal dominant connective tissue disease associated with significant morbidity and mortality due to progressive dilatation of the thoracic aorta which can lead to aortic rupture. Survival from an aortic rupture is predicated on immediate organized and goal directed care by both surgical and anesthesia teams. This case highlights how coordinated care from a cardiac operating room team, including early preparation of autologous blood products, expeditious placement of intravascular access for rapid high volume transfusion, and intentional communication between anesthesia, perfusion, surgery and nursing during the resuscitation in the OR, can all lead to an improved outcome.

1. Introduction

Marfan syndrome (MS) is an autosomal dominant connective tissue disease caused by a mutation in the FBN1 gene [1,2]. MS is associated with progressive dilation of the aortic root and thoracic aorta that can lead to a host of aortic syndromes including aneurysm, dissection and rupture [3]. Much like the trauma setting, thoracic aortic rupture carries a high mortality, which approaches 97 % if free rupture occurs [4,5]. Surviving aortic rupture is predicated on immediate organized and goal directed care by both surgical and anesthesia teams [4,6].

In the setting of aortic rupture, several systems and technical factors can all potentially protect against mortality. Early utilization of massive transfusion protocols are critical to maintain blood volume, clotting capability and tissue oxygen delivery until bleeding is controlled. The immediate availability of circulatory support, including cardiopulmonary bypass (CPB) and deep hypothermic circulatory arrest (DHCA), is another vital component to bridging to definitive aortic repair [7]. These protective strategies can require an immense amount of organization and preparedness.

We present a case of a complex MS patient who survived a thoracic aortic rupture at our institution. Written consent for the publication of this case was provided by the patient.

* Corresponding author.
E-mail addresses: talonljones@gmail.com (T. Jones), joel.price@vch.ca (J. Price), sean.mclean@vch.ca (S.R. McLean).

https://doi.org/10.1016/j.heliyon.2024.e25235
Received 23 November 2023; Received in revised form 22 January 2024; Accepted 23 January 2024
Available online 28 January 2024

Glossary of terms

MS	Marfan Syndrome
OR	operating room
CPB	cardiopulmonary bypass
DHCA	deep, hypothermic circulatory arrest
pRBC	packed red blood cells
FFP	fresh frozen plasma
ACLS	Advanced cardiac life support
PEA	pulseless electrical activity
CSICU	cardiac surgery intensive care unit
POD	postoperative day

2. Case description

A 22-year-old male with a history of MS and previous aortic valve-sparing root replacement 3 years prior presented to hospital emergency department complaining of severe back pain and dizziness. An urgent CT was performed (Fig. 1) which demonstrated severe pectus excavatum, and previous aortic root replacement. There was an ascending aortic pseudoaneurysm and an accompanying para-aortic hematoma originating from the previous distal suture line and extending into the aortic arch. These findings were consistent with a contained aortic arch rupture. Blood pressure control was initiated in the emergency department with a total of 70mg of intravenous labetalol while preparations were made for transport to the operating room (OR). Initially the patient's systolic blood pressure was 105 mmHg, heart rate 75 beats per minute, and his respiratory status was stable on 2 L of oxygen via nasal prongs, however, his clinical course began to worsen during transport to the OR with signs of respiratory distress including rapid shallow breathing and progressive central chest pain. This was likely due to increasing left mainstem bronchial compression from the expanding, contained aortic rupture. Upon arrival in the OR, high flow oxygen was applied while large bore intravenous access and intra-arterial monitoring were established and a rapid transfuser was primed. 8 units of cross matched packed red blood cells (pRBC), 4 units of fresh frozen plasma (FFP) and 2 pooled platelets were made immediately available. Perfusion support was in the room at the time of induction of anesthesia with a cardiopulmonary bypass (CPB) circuit primed and in the room. Induction of anesthesia was performed and the airway was secured uneventfully with negligible changes in hemodynamics. A right internal jugular multi access lumen catheter, right sided dual lumen central venous catheter, two additional 14 gauge peripheral IVs, and bilateral upper extremity art lines were obtained.

During preparation for surgery, the patients legs were elevated to accommodate skin preparation with chlorhexidine. Concomitant with the leg raise, there was an acute drop in mean arterial pressure from 81 to 25 mm Hg. The heart rate decreased to 30 and the ECG revealed a junctional rhythm. A code blue for pulseless electrical activity (PEA) was announced and chest compressions were commenced with the presumed diagnosis being complete aortic rupture and accompanying massive hemorrhage. Advanced cardiac life support (ACLS) for PEA was initiated and a massive transfusion protocol was initiated. Personnel were recruited to exclusively manage both ACLS and blood product management. Two cell saver units were used during this time.

As the patient had a significant pectus excavatum and had received a prior sternotomy, direct and expeditious control of the aorta was not feasible. With ACLS for PEA ongoing, heparin 30000 units IV was administered, and the cardiac surgeon immediately

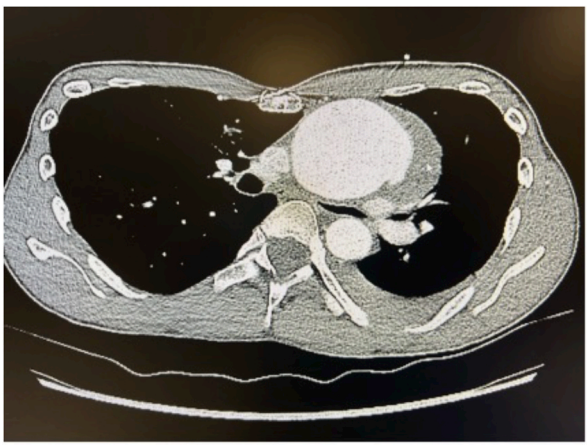


Fig. 1. CT Angiography demonstrating severe pectus excavatum, sternal wire from previous sternotomy, contained ascending aortarupture and associated left mainstem bronchus compression and small left pleural effusion.

performed a right femoral cutdown to establish femoral arterial and venous access for CPB via 19 Fr (arterial) and 27 Fr (venous) cannulas. A transesophageal echo probe, a pulmonary artery catheter, and bilateral brachial arterial lines were inserted during the resuscitation to evaluate cardiac activity and volume status. Throughout the salvage resuscitation, and prior to initiation of CPB, high quality chest compressions ensured adequate blood pressure (>50 mmHg) and an end tidal CO_2 greater than 20 cmH20, as per ACLS guidelines. ACLS was performed for 20 minutes prior to successful initiation of femoral CPB, and by this point a total of 29 pRBCs, 8 units of FFP, 8 g of fibrinogen and 3 units of platelets had already been transfused. The central venous pressure ranged between 40 and 50 mmHg during the resuscitation, likely a result of tamponade effect in the mediastinum [1]. Upon initiation of CPB, cooling towards deep hypothermia at 18 °C was initiated.

Once the redo sternotomy and exposure of the aorta was completed on CPB, a rupture of the pseudoaneurysm at the junction of the prior Dacron graft and the distal ascending aorta was encountered. Digital control of the hemorrhage was obtained and the cooling was completed. After achieving a core temp of 18 °C, DHCA was initiated and the aorta was resected from the previous graft into zone 2. A cannula was inserted directly to the brachiocephalic artery and antegrade cerebral perfusion was administered. Standard aortic arch reconstruction was performed including distal anastomosis in zone 2, interposition grafts to the brachiocephalic and left carotid arteries and proximal anastomosis directly to the previous Dacron graft.

Total surgical time was 10.5 hours, with 243 minutes of aortic cross clamp, 357 minutes on CPB, 102 minutes of total DHCA, and 64 minutes of circulatory arrest with antegrade cerebral perfusion. Thromboelastography and coagulation measurements (ACT, INR, fibrinogen, PTT, CBC) were taken serially to guide blood product transfusion, and a total of 54 pRBC, 22 FFP, 13 platelets, 20g of fibrinogen, and 5mg of activated factor VII were administered. Due to ongoing coagulopathy and bleeding, the patient's chest was not closed and he was transported to the cardiac surgery intensive care unit (CSICU) under deep sedation on a norepinephrine infusion of 8 mcg/min IV. On post operative day (POD) 1 the patient underwent re-evaluation, hematoma evacuation and closure of sternotomy.

The patient's neurological prognosis was extremely guarded due to the prolonged resuscitation and elevated central venous pressure during chest compressions prior to circulatory arrest. Neuroimaging on POD1 revealed acute supra and infratentorial infarcts bilaterally, as well as loss of grey-white differentiation (Fig. 2). However, on POD 6, neurological function slowly began to improve. A tracheostomy was performed, and the patient continued to improve neurologically to the point that he was alert, responsive and communicating by POD12. There was delayed recovery of motor function to his lower extremities, and he was found to have suffered a spinal cord infarction from T-10 to conus. Other complications encountered over his one month ICU stay included: a ventilator associated-pneumonia; demyelinating peripheral neuropathy; and a para-aortic graft collection that was treated conservatively. He was transferred to a local rehabilitation center two months after his initial surgery and remained there for an additional three months of inpatient rehab. At the time of discharge, he was performing activities of daily living independently with appropriate aides, and ambulating independently using a four wheeled walker.

3. Discussion

Aortic arch ruptures carry significant mortality especially in the context of free non-contained proximal ruptures, with over 90 % mortality reported [8,9]. Early preparation, team-based goal directed care, and prompt transition to CPB and DHCA, were all imperative measures in keeping this patient alive and offering a chance for a good outcome.

With respect to preparation for massive transfusion, standardized protocols for aortic arch rupture are rare, given that the majority of ruptures are seen in the trauma setting and do not survive. In this case, known pseudoaneurysm and proximal rupture warranted extensive precautionary strategies including: priming rapid transfusion devices and cell savers; early massive transfusion protocol

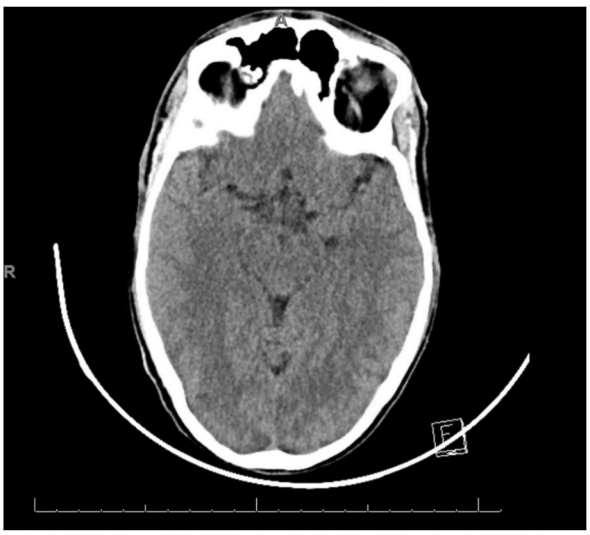


Fig. 2. CT Head- Postoperative day 3, demonstrates loss of grey-white differentiation consistent with severe hypoxic brain injury.

activation; and dedicating additional resuscitation team members for blood product checking and transfusion. Perfusion services were notified early and there was a primed CPB machine in the room.

Throughout the resuscitation, sequential landmarks for continuation were communicated to keep the team focussed and to delineate where ongoing resuscitation could be halted due to futility. After the rupture, establishing effective chest compressions was reassuring for end organ perfusion and oxygen delivery. Subsequently, initiating CPB facilitated blood pressure support and core temperature cooling to 18 °C to protect the brain and spinal cord. Finally, DHCA was used to facilitate surgical exposure for definitive hemorrhage control and selective cerebral perfusion after redo sternotomy.

Critical features in this case included: concerted and organized care from a comprehensive cardiac OR team; early preparation of autologous blood products; expeditious placement of intravascular access for rapid high volume transfusion, and intentional communication between anesthesia, perfusion, surgery and nursing during the resuscitation in the OR. Similar algorithmic approaches have been demonstrated to improve outcomes in the ruptured abdominal aortic aneurysm population [10]. Persistent effort and care was provided after aortic rupture occurred, but balanced with well-defined sequential resuscitation goals. We believe that these resuscitation tenants can be replicated in similar situations where the threat of catastrophic aortic rupture is high, and hopefully improve outcomes.

Declaration

Written informed consent for the publication of case details and images was provided by the patient.

Data availability

No data was used for the research described in the article.

CRediT authorship contribution statement

Talon Jones: Writing – original draft. **Joel Price:** Writing – review & editing, Supervision. **Sean R. McLean:** Writing – review & editing, Writing – original draft, Supervision.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

[1] M.J. Roman, R.B. Devereux, Aortic dissection risk in marfan syndrome, J. Am. Coll. Cardiol. 75 (8) (2020) 854–856.
[2] E.M. Isselbacher, O. Preventza, J. Hamilton Black, et al., ACC/AHA guideline for the diagnosis and management of aortic disease: a report of the American heart association/American college of cardiology joint committee on clinical practice guidelines, Circulation 146 (24) (2022) e334–e482, 2022.
[3] U. Benedetto, G. Melina, J.J. Takkenberg, et al., Surgical management of aortic root disease in Marfan syndrome: a systematic review and meta-analysis, Heart 97 (12) (2011) 955–958.
[4] N. Boucher, H. Dreksler, J. Hooper, et al., Anaesthesia for vascular emergencies - a state of the art review, Anaesthesia 78 (2) (2023) 236–246.
[5] G. Johansson, U. Markström, J. Swedenborg, Ruptured thoracic aortic aneurysms: a study of incidence and mortality rates, J. Vasc. Surg. 21 (6) (1995) 985–988.
[6] T. Fukui, Management of acute aortic dissection and thoracic aortic rupture, J Intensive Care 1 (6:15) (2018).
[7] N. Hynes, T. Mastracci, Building a complex thoracic aortic program from the ground up: what does it take, EndoVasc Today 18 (11) (2019) 58–64.
[8] D. Sidloff, E. Choke, P. Stather, et al., Mortality from thoracic aortic diseases and associations with cardiovascular risk factors, Circulation 130 (25) (2014) 2287–2294.
[9] R.S. Bonser, A.M. Ranasinghe, M. Loubani, et al., Evidence, lack of evidence, controversy, and debate in the provision and performance of the surgery of acute type A aortic dissection, J. Am. Coll. Cardiol. 58 (24) (2011) 2455–2474.
[10] E.L. Chaikof, DalmanRL, M.K. Eskandari, et al., The Society for Vascular Surgery practice guidelines on the care of patients with an abdominal aortic aneurysm, J. Vasc. Surg. 67 (1) (2018).