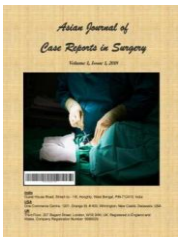

Asian Journal of Case Reports in Surgery

2(1): 6-12, 2019; Article no.AJCRS.45746

Surgical Treatment of a Ruptured Pseudoaneurysm of the Descending Thoracic Aorta Associated with Takayasu's Arteritis - A Case Report of a 9-year-old Boy

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Authors' contributions

This work was carried out in collaboration between all authors. Author QTT designed the study, and reviewed the drafted manuscript. Author TKVL was the main surgeon, managed the analyses of the study. Authors VNL and VPD are senior surgeon, followed up the patient during the period. Author HMP is an intensivist, followed up the patients during the period. Author TBH wrote the first draft of the manuscript, wrote the protocol, and managed the literature searches. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/AJCRS/2019/45746
Editor(s):
(1) Dr. Guven Aslan, Professor, Department of Urology, Dokuz Eylul University School of Medicine, Turkey.
Reviewers:
(1) Eyo E. Ekpe, University of Uyo, Nigeria.
(2) Suraj Wasudeo Nagre, Grant Medical College, India.
(3) B. Ngo Nonga, University of Yaoundé I, Cameroon.
(4) P. K. Hota, NTR University of Health Sciences, India.
Complete Peer review History: http://www.sciencedomain.org/review-history/28131

Case Study

Received 06 October 2018
Accepted 12 December 2018
Published 04 January 2019

ABSTRACT

Takayasu's arteritis (TA) is a chronic inflammatory disease of the aorta and its major branches. Aortic aneurysm is a severe complication of TA, with an estimated mortality rate of 50% in patients that undergo surgery for a rupture [1]. We herein reviewed the clinical, paraclinical and operation characteristics of a 9-year-old male patient, who had a ruptured pseudoaneurysm of the thoracic aorta due to TA, and was successfully treated by open emergency surgery with cardiopulmonary bypass support.

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Keywords: Takayasu's arteritis; thoracic pseudoaneurysms.

ABBREVIATIONS

TA: Takayasu's arteritis; CT scan: Computed Tomography scan, HLA: Human leukocyte antigens.

1. INTRODUCTION

Takayasu's arteritis (TA) is an idiopathic chronic vasculitis of large vessels and their main branches that rarely affects children. The inflammation results in varying degrees of stenosis, occlusion or dilation of the involved vessels. The aetiology and precise pathogenesis of TA remain unclear but much has been learnt about the disease since 1908, after the first report by Mikito Takayasu, a Japanese ophthalmologist [2]. Arterial stenosis was one of the most common lesions with about 90% presented in the TA patients that were mentioned in global literature. Aneurysms and pseudoaneurysm conditions were less common with an incidence of 25% [3]. However, they had a high risk of rupture with high mortality rates of up to 50% [1]. We herein report a case of a 9-year-old male patient who underwent open repaired surgery for a ruptured thoracic pseudoaneurysm associated with Takayasu arteritis with cardiopulmonary bypass support.

2. CASE PRESENTATION

2.1 Past Medical History

A 9-year-old male patient, who had an unremarkable medical history, presented symptoms including fatigue, cough, mid-grade fever and persistent left chest pain radiating to his back. The symptoms lasted for one week before he was transferred to a local hospital due to a condition of hemoptysis. The plain chest X-ray revealed an abnormal shadow on the middle of his left chest, and laboratory examination showed an increase of white blood cells with 15 K/µL. The initial diagnosis was caseous pneumonia differentiated from pulmonary tuberculosis infection.

Four hours later, the patient presented a sudden onset of left chest pain, dyspnea and early signs of hypovolemic shock. A computer tomography with contrast was done immediately, the figures revealed a ruptured pseudoaneurysms of the descending thoracic aorta. After receiving a blood transfusion, the patient was transferred to Cho Ray Hospital 30 minutes later.

2.2 Physical Findings

- Vital signs: Pulse rate 130 beats per minute, body temperature 37 degrees C, respiratory rate 30 breaths per minute, blood pressure on left arm 125/85 mmHg, right arm 120/90 mmHg, both legs could not measure blood pressure, femoral pulse was absent.
- The patient was fatigued, with pallor skin.
- Examination of the chest revealed tachycardia, dullness and diminished breath sounds on the left side.

2.3 Laboratory Data

Red blood cell count: 3.69 T/L, Haematocrite 28.3%, Hemoglobin 89 g/L, White blood cell count 16 G/L, Neutrophyle 78.7%, Platelet count 233 G/L, C-reactive protein 30.8 mg/L, Erythrocyte sedimentation rate (ESR) rose to 44mm per hour and 84 mm per 2 hours.

2.4 Imaging Diagnosis

- **Chest x-ray**: Large pleural effusion, the mediastinal structures were shifted mediastinal to the right (Fig. 1).

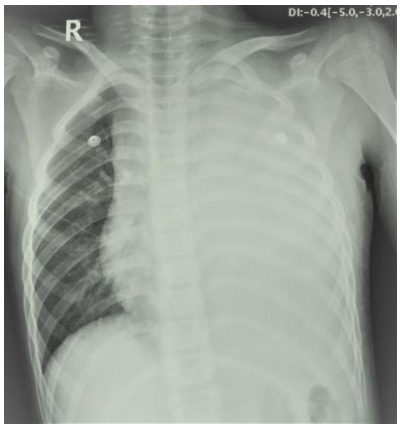

Fig. 1. The plain chest x-ray shows a large pleural effusion

- Chest CT scan with contrast (Fig. 2) showed a large haemothorax. The descending thoracic aorta presented a pseudoaneurysm of 8 centimeters in diameter, the thoracic aorta below the pseudoaneurysm presented a severe

stenosis of about 0.7 centimeters in diameter and 3 centimeters in length. The superial mesenteric artery also revealed stenosis.

- Echography: Besides the pseudoaneuryms and stenosis of the thoracic aorta shown by the CT scan, echography results found a moderate pericardial effusion, aneurysms of the left common carotid artery, atherosclerosis and mild stenosis of the iliac and femoral artery on both sides.

2.5 Treatment

Our surgical team decided that the patient was a candidate for emergency surgery. A sternotomy was performed. We exposed the entire aorta arch and descending aorta just above the pseudoaneurysm, set up circulation extracorporelle via the ascending aorta, right femoral artery, superior vena cava and inferior vena cava. The body temperature was reduced to 33 degrees C. After opening the pleural cavity, we saw a lot of blood and blood clots estimated at about 1.5 liters. The pseudoaneurysms at the descending thoracic aorta were about 12x10 cm in size. The aorta just above the diaphragm had severe stenosis (Fig. 3).

We clamped the aorta above the neck of the pseudoaneurysms as well as below the stenosis position. A Dacron graft was used to repair the aorta (Fig. 4). The aortic cross-clamping time was 45 minutes, and the cardiopulmonary bypass time was 120 minutes.

The postoperative course was uneventful. The patient was treated with antibiotics, pain relief and corticoid therapy. The histopathological figures showed necrosis of the aorta wall and infiltration of lymphocytes, plasmacytes and neutrophils. A CT scan in the postoperative period confirmed that the entire descending thoracic aorta was repaired successfully (Fig. 5). The postoperative plain chest x-ray showed a shadow on the left side, which was the remnants of the pseudoaneurysm cover, and the left lung had expanded completely (Fig. 5).

The patient was discharged on the 16th day after the operation. He was transferred to a pediatric hospital for follow up and to take care of the Takayasu arteritis condition.

3. DISCUSSION

Takayasu's arteritis (TA) is a large vessel systemic granulomatous vasculitis primarily involving the aorta and its major branches. It is reported all around the world. However, it is more common in South-East Asia, Africa and South America [4]. This disease is the most common

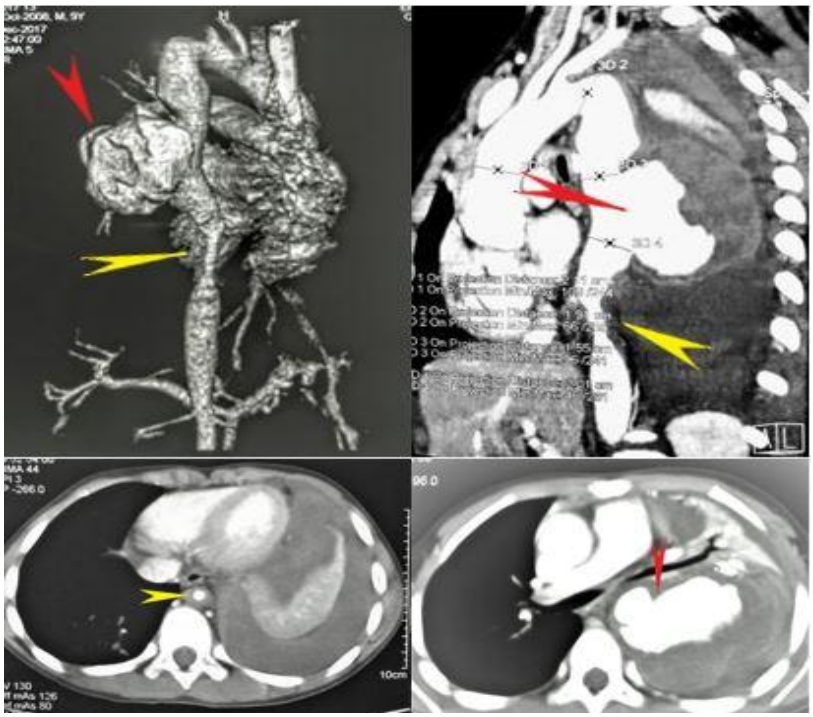

Fig. 2. CT scan figures show a ruptured pseudoaneurysms of 8 centimeters in diameter (red arrow) and a 3cm stenosis (yellow arrow) of the descending thoracic aorta

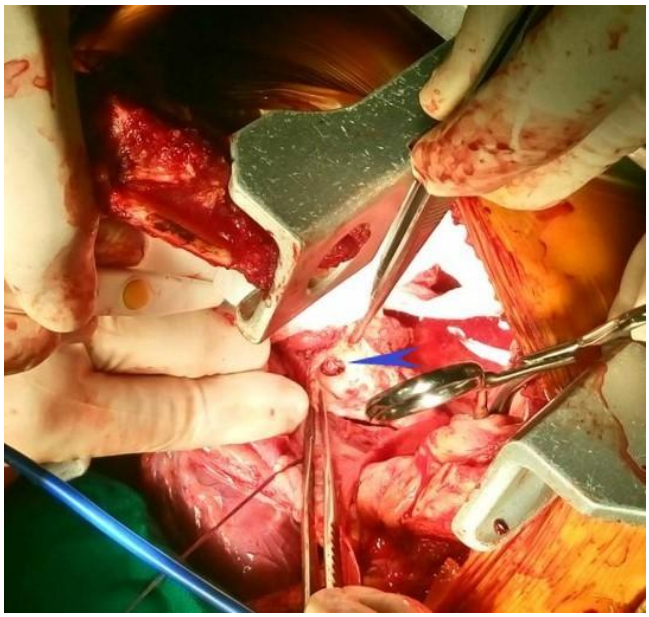

Fig. 3. Severe stenosis of the descending thoracic aorta just above the diaphragm (blue arrow)

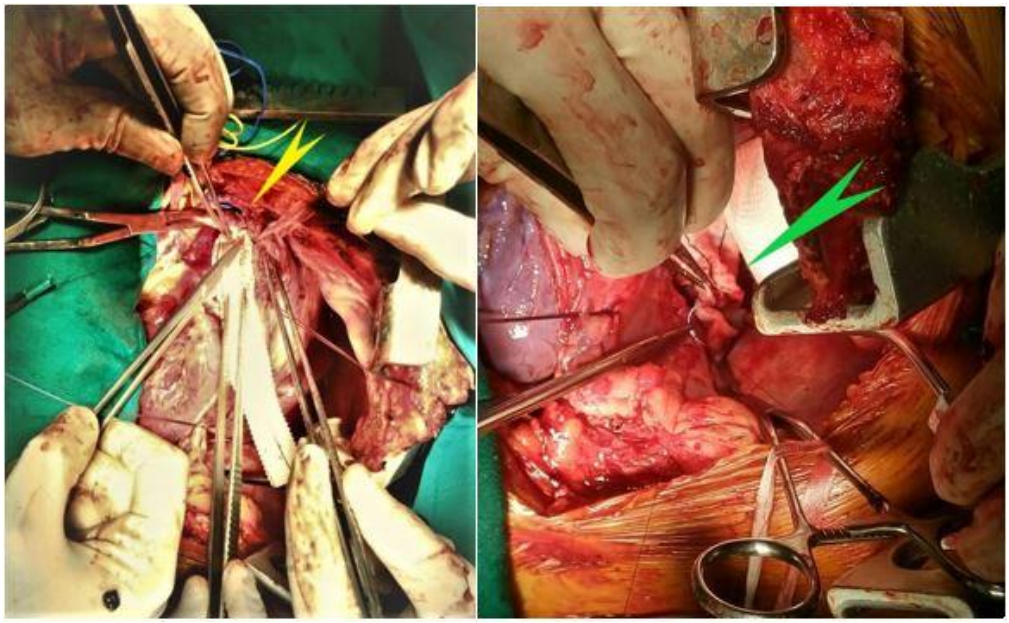

Fig. 4. Making a connection between the upper (yellow arrow) and lower part (green arrow) of the descending thoracic aorta and Dacron graft

cause of renohypertension in Asian children [5]. According to a systemic review in Japan during the period from 1982 to 1984, there were 2600 patients with TA [6]. In the United States of America, the incidence was estimated at 2.6 per million population/year [7].

Takayasu's arteritis is reported in young adults in the second and third decades of life. The onset of symptoms may be earlier, including in childhood, but rarely in infancy. The female:male ratio was about 9:1 according to reports from Japan [6]. In Vietnam, according to a report from the Ho Chi Minh city Children's Hospital, the female:male ratio was about 1.63:1 [8]. The exact pathogenesis of TA is unclear. Cell-mediated immunity has an important role in the mechanisms of arteritis. The genetic factors which relate to TA have been extensively studied. The research from Japan showed that

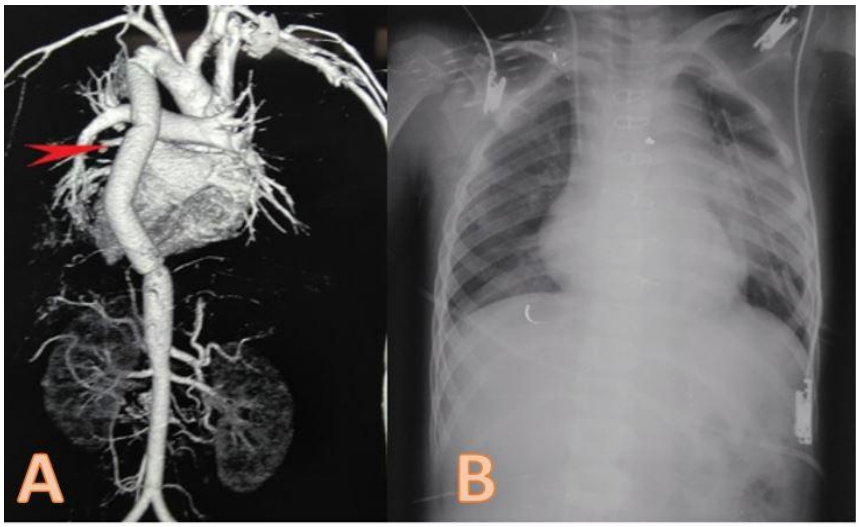

Fig. 5. The CT scan confirmed that the entire descending thoracic aorta was replaced by the Dacron tube (A). The plain chest x-ray showed a shadow which was the remnants of the pseudoaneurysm cover (B).

Table 1. European league against rheumatism (EULAR)/ Pediatric Rheumatology International Trials Organization (PRINTO)/ Pediatric Rheumatology European Society (PRES) classification criteria of Takayasu's arteritis and additional criteria

Mandatory criteria Angiographic abnormality	Angiography (conventional, CT, and MRI) of the aorta, its main branches or pulmonary arteries	Aneurysm/ dilatation, narrowing, occlusion, or thickened arterial wall, not due to any other causes
Additional criteria (need one of five)	1. Pulse deficit or claudication	- Lost/decreased/unequal peripheral artery pulse - Symptoms of claudication: focal muscle pain induced by physical activity
	2. Blood pressure discrepancy	Discrepancy of four limb systolic blood pressure >10 mmHg in any limb
	3. Bruits	Audible murmurs or palpable thrills over large arteries
	4. Hypertension	Systolic/diastolic blood pressure >95th centile for height
	5. Acute phase reactant	Erythrocyte sedimentation rate >20 mm per hour or C-reactive protein above normal

HLA B-52 and DR-2 are associated with TA. In Korea, there has also been a report of HLA B-52 and B-5 association [9]. Some recent studies show that there was a close association between Tuberculosis and Takayasu. Furthermore, the epidemiological regions of Tuberculosis and Takayasu were similar. In Vietnam, a study conducted by the Children's Hospital in 2003 found that 23.8% of Takayasu patients had Tuberculosis or someone in their family had Tuberculosis [8].

To make a diagnosis of Takayasu's arteritis on this patient, we based it on clinical criteria. Several criteria of differing sensitivity and specificity have been offered. We based our diagnosis on the *European League Against Rheumatism (EULAR)/Pediatric Rheumatology International Trials Organization (PRINTO)/Pediatric Rheumatology European Society (PRES)* which validated the TA criteria for children [10] (Table 1). Diagnosis requires the presence of mandatory criteria and at least one additional criteria. For this patient, the figures of the CT scan showed narrowing of the aorta, iliac and femoral artery, and it also had a pseudoaneurysm. We also had 3 of 5 additional features, which were (1) loss of peripheral artery

pulse (femoral pulse was absent),(2) discrepancy of four limb systolic blood pressure >10 mmHg in any limb (blood pressure on left arm 125/85 mmHg, right arm 120/90 mmHg, while the blood pressure in both legs could not be measured) and (5) Erythrocyte sedimentation rate >20 mm per hour or C-reactive protein above normal (ESR 44mm per hour and CRP 30.8 mg/L).

The diagnosis is difficult to make during the acute early phase, because patients show symptoms of systemic inflammation such as fever, fatigue, headache, muscle pain, arthralgia, and weight loss. Laboratory findings such as erythrocyte sedimentation rate and C-reactive protein show an increase. All of them are nonspecific. Therefore, the disease often goes unrecognised in this phase. This patient presented symptoms including fatigue, cough, mid-grade fever and persistent left chest pain radiating to his back. In addition, the plain chest x-ray showed a shadow on the left side of the thorax. All of the clinical findings were similar to pneumonia or pulmonary Tuberculosis. For this reason, the local hospital made the mistake on initial diagnosis. Ruptured pseudoaneurysm was not diagnosed until the boy had symptoms of pseudoaneurysm rupture with violent chest pain, and hypovolemic shock, which were confirmed by the CT scan.

Aortic aneurysm, as well as pseudoaneurysm, are potentially life-threatening risk factors, with an estimated mortality rate of 80%. The progress of this ruptured pseudoaneurysm was extremely quick because of the presence of a stenosis just below, which caused high pressure on the wall of pseudoaneurysm.

After performing an open repair of the thoracic aorta, postoperative steroid therapy was the primary means of controlling the degree of inflammation. The boy had many problems such as aneurysms of the left common carotid artery, atherosclerosis and mild stenosis of iliac and femoral artery on both sides. All of these need continuous follow up.

4. CONCLUSION

Takayasu’s arteritis is a rare disease in children. To date, with the development of non-invasive diagnostic imaging tools, the final diagnosis can be made earlier. With the development of a new generation of immunosuppressive drugs combined with classical steroids as well as more advances in surgical technology and perioperative care, the prognosis of TA has improved. However, despite these developments, the management of TA in children still has many challenges because the diagnosis is still often late and currently there is no sensitive and specific gold standard for the monitoring of inflammatory diseases. Treatment is essential to prevent the insidious course and vascular damage from this disease, including stenosis, occlusive lesions, aneurysms with dangerous complications and a high rate of mortality.

CONSENT AND ETHICAL APPROVAL

As per university standard guidelines, participant consent and ethical approval has been collected and retained by the authors.

ACKNOWLEDGEMENT

We thank our colleagues from Cardiovascular Centre of Cho Ray Hospital who provided insight and expertise in taking care of this patient.

We would also like to show our gratitude to Ms. Elanie Marks (World Health Organization) and Mr. Michael Schademan (JOLO English Centre) for comments and revisions that greatly improved the manuscript.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sciencedomain.org/review-history/28131