

Case study

Surgical treatment of ruptured pseudoaneurysm

of the descending thoracic aorta associated with Takayasu's arteritis A case report of a 9-year-old boy

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.

Key word: *Takayasu's arteritis, thoracic pseudoaneurysms.*

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 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.

CASE PRESENTATION

Past medical history

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

36 Four hours later, the patient presented a sudden onset of left chest pain, dyspnea and early
37 signs of hypovolemic shock. A computer tomography with contrast was indicated
38 immediately, the figures revealing a ruptured pseudoaneurysms of the descending thoracic
39 aorta. After being get blood transfusion, the patient was transferred to Cho Ray Hospital 30
40 minutes later

41 **Physical Findings**

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

48 **Laboratory data**

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

52 **Imaging diagnosis**

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



Figure 1: The plain chest x ray shows a large pleural effusion

- **Chest CT scan with contrast** (Figure 2): Descending thoracic aorta presented a pseudoaneurysm with 8 centimeters in diameter, thoracic aorta below the pseudoaneurysm presented a severe stenosis about 0.7 centimeters in diameter lasting for 3 centimeters, large haemothorax, superior mesenteric artery also revealed stenosis.

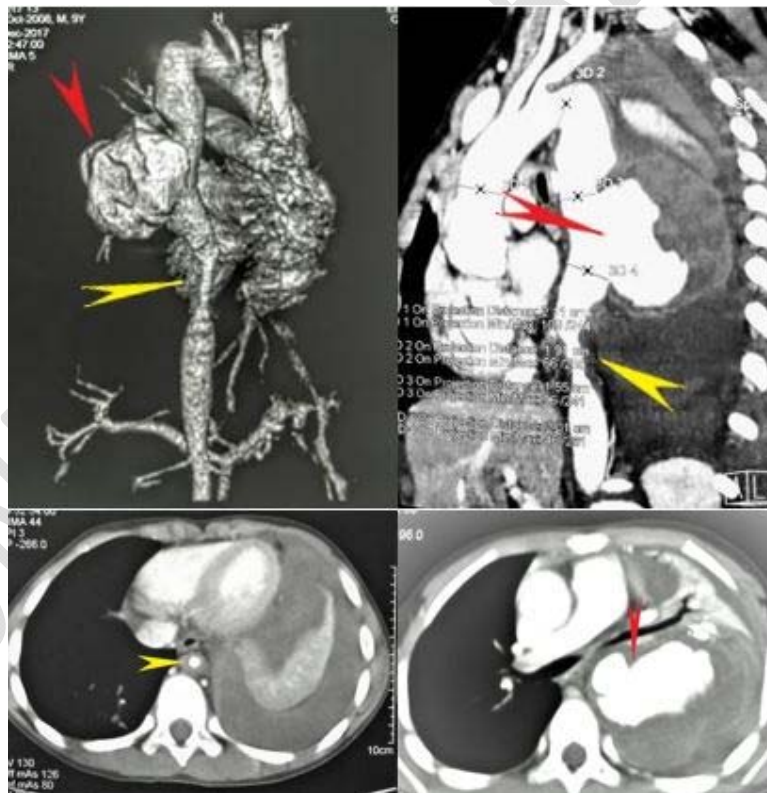
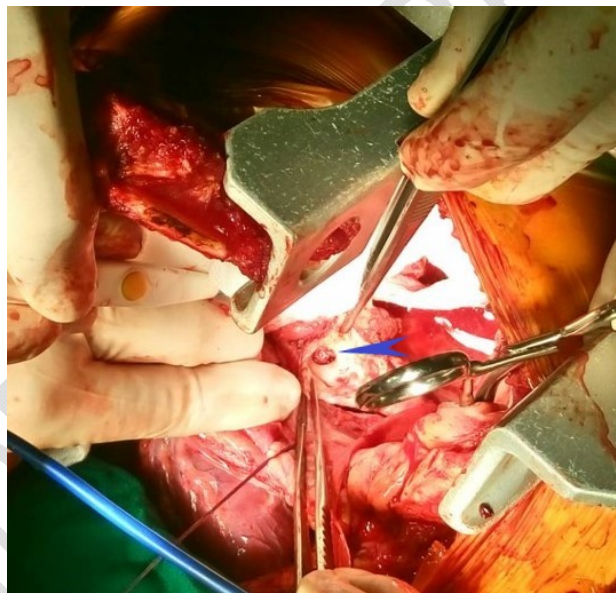


Figure 2: CT scan figures show a ruptured pseudoaneurysms with 8 centimeters in diameter (red arrow) and a 3cm stenosis (yellow arrow) of descending thoracic aorta

64 - **Echography:** Beside the pseudoaneurysms and stenosis of thoracic aorta shown by the CT
65 scan, echography results found a moderate pericardial effusion, aneurysms of the left
66 common carotid artery, atherosclerosis and mild stenosis of the iliac and femoral artery on
67 both sides.

68 **Treatment**

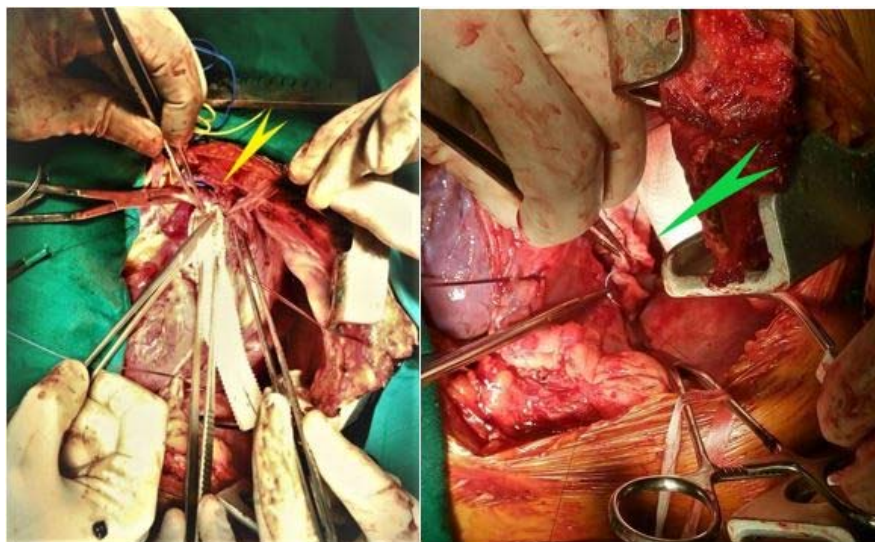
69 Our surgical team decided that the patient was a candidate for emergency surgery. A
70 sternotomy was performed, we exposed the entire aorta arch and descending aorta just above
71 the pseudoaneurysm, set up circulation extracorporelle via the ascending aorta, right femoral
72 artery, superior vena cava and inferior vena cava. The body temperature was reduced to 33
73 degrees C. After opening the pleural cavity, we saw a lot of blood and blood clots estimated
74 at about 1.5 liters. The pseudoaneurysms at the descending thoracic aorta were about
75 12x10cm in size. The aorta just above the diaphragm had severe stenosis (Figure 3).



76

77 *Figure 3: Severe stenosis in descending thoracic aorta just above the diaphragm (blue arrow)*

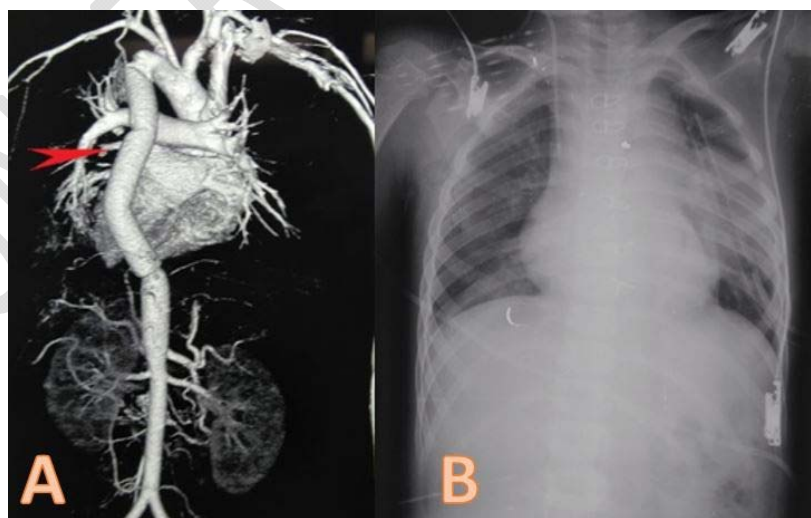
78 We clamped the aorta above the neck of the pseudoaneurysms as well as below the stenosis
79 position. A Dacron graft was used to repair the aorta (Figure 4). The aortic cross-clamping
80 time was 45 minutes, cardiopulmonary bypass time was 120 minutes.



81

82 *Figure 4: Making a connection between the upper (yellow arrow) and lower part (green arrow) of*
 83 *the descending thoracic aorta and Dacron graft*

84 The postoperative course was uneventful, the patient was treated by antibiotic, pain relief and
 85 corticoid therapy. The histopathological figures showed necrosis of the aorta wall and the
 86 infiltration of lymphocytes, plasmacytes and neutrophils. We indicated a CT scan in the
 87 postoperative period. The figures of the CT scan confirmed that the entire descending
 88 thoracic aorta was repaired successfully (Figure 5). The postoperative plain chest x-ray
 89 showed a shadow in the left side, which was the remnants of pseudoaneurysm cover, the left
 90 lung had expanded completely (Figure 5) .



91

92 *Figure 5: CT scan confirmed that the entire descending thoracic aorta was replaced by Dacron tube*
 93 *(A), Plain chest x ray showed a shadow which was the remnants of the pseudoaneurysm cover (B).*

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

96 **DISCUSSION**

97 Takayasu's arteritis (TA) is a large vessel systemic granulomatous vasculitis primarily
98 involving the aorta and its major branches. It is reported all around the world. However, it is
99 more common in south-east Asia, Africa and South America [4]. This disease is the most
100 common cause of renohypertension in Asian children [5]. According to a systemic review in
101 Japan during the period from 1982 to 1984, there were 2600 patients with TA [6]. In United
102 States of America, the incidence was estimated at 2.6 per million population/year [7].

103 Takayasu's arteritis is practically reported in young adults in the second and third decades of
104 life. The onset of symptoms may be earlier, including in childhood, but rarely in infancy. The
105 female:male ratio was about 9:1 in some reports from Japan [6]. In Vietnam, according to a
106 report of Ho Chi Minh city Children's Hospital, the female:male ratio was about 1.63:1 [8].
107 The exact pathogenesis of TA has been unclear, cell-mediated immunity has an important
108 role in the mechanisms of arteritis. Genetic factor, which relates to TA, has been extensively
109 studied. The research from Japan showed that HLA B-52 and DR-2 are associated with TA,
110 Korea also had a report of HLA B-52 and B-5 association [9]. Some recent studies show that
111 there was a close association between Tuberculosis and Takayasu. Furthermore, the
112 epidemiological regions of Tuberculosis and Takayasu were similar. In Vietnam, a study
113 conducted by the Children's Hospital in 2003 found that 23.8% of Takayasu patients had
114 Tuberculosis disease or their family had Tuberculosis [8].

115 To make a diagnosis of Takayasu's arteritis on this patient, we based it on clinical criteria.
116 Several criteria of differing sensitivity and specificity have been offered. We based on the
117 *European League Against Rheumatism (EULAR)/Pediatric Rheumatology International*
118 *Trials Organization (PRINTO)/Pediatric Rheumatology European Society (PRES)* validated
119 the TA criteria for childhood [10] (Table 1). Diagnosis requires the presence of mandatory
120 criteria and at least one additional criteria. On this patient, the figures of the CT scan showed
121 narrowing of the aorta, iliac and femoral artery, it also had a pseudoaneurysm. We also had 3
122 of 5 additional features, those were (1) lost peripheral artery pulse (femoral pulse was
123 absent), (2) discrepancy of four limb systolic blood pressure >10 mmHg in any limb (blood
124 pressure on left arm: 125/85 mmHg, right arm 120/90 mmHg, while both of legs could not

measure blood pressure) and (5) Erythrocyte sedimentation rate >20 mm per hour or C-reactive protein above normal (ESR 44mm per hour and CRP 30.8mg/L).

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	Angiography (conventional, CT, and MRI) of the the aorta, its main branches or pulmonary arteries	Aneurysm/ dilatation, narrowing, occlusion, or thickened arterial wall, not due to any other causes
Angiographic abnormality		
	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
Additinal criteria (need one of five)	3. Bruits	Audible murmurs or palpable thrills over large arteries
	4. Hypertension	Systolic/diastolic blood pressure >95 th centile for height
	5. Acute phase reactant	Erythrocyte sedimentation rate >20 mm per hour or C-reactive protein above normal.

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. The 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 a pneumonia or pulmonary Tuberculosis. That was the reason why the local hospital made the mistake on initial diagnosis. Until the boy had the symptom of pseudoaneurysm rupture with violent chest pain, hypovolemic shock, the CT scan had been indicated to confirm.

Aortic aneurysm as well as pseudoaneurysm are a potentially life-threatening risk factor, with the estimated mortality to be 80% and 50% in patients who arrive at hospital and who undergo surgery for a rupture, respectively. The progress of ruptured pseudoaneurysm was extremely quick because of the presence of stenosis just below which made high pressure on the wall of pseudoaneurysm.

After performing open repair of the thoracic aorta, postoperative steroid therapy was a main point to control 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 them need to be followed up continuously.

CONCLUSION

Takayasu's arteritis is a rare disease in children. To date, with the development of non-invasive diagnostic imaging tools, the final diagnosis had been 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 diagnosis is still often late and currently there is no sensitive and specific gold standard for the monitoring of inflammatory disease. Treatment is essential to prevent the insidious course and vascular damage: stenosis, occlusive lesions, aneurysms with dangerous complications and high rate of mortality.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

ABBREVIATIONS

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

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