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Case Report

Ascending Aortic Aneurysm Caused by Takayasu`s Arteritis: A Case Report

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Abstract

Takayasu arteritis (TA) is a rare systemic vasculitis primarily affecting large and medium-sized arteries. It predominantly presents in young femalesa and can lead to significant complications, including aortic aneurysms. We present a case of a 42-year-old male with a history of TA and multiple arterial aneurysms, including an ascending aortic aneurysm measuring 5.7 cm. After routine follow-up and imaging revealed the increasing size of the aneurysm, surgical intervention was deemed necessary. The patient underwent successful surgical repair through median sternotomy and aneurysm plication on cardiopulmonary bypass

KEYWORDS

Cardiac surgery; Aortic surgery; Takayasu`s Arteritis

Introduction

Takayasu arteritis (TA) is an idiopathic, rare, systemic disease that affects large and medium-sized arteries and their branches, the most commonly involved vessel being the aorta and its branches [1-3]. It occurs most commonly in young adults and predominantly in females, usually presenting in 2nd and 3rd decades of life, but can also affect children. The disease is more prevalent in Asians, although it has been reported from all parts of the world, and its incidence ranges from 0.3-3.3 million per year [4]. Initially, it presents with non-specific, constitutional symptoms like malaise, fever, anorexia, and weight loss, and about 10% of patients are asymptomatic [3]. Other features at presentation include neurologic manifestations, hypertension, differences in blood pressure between extremities, carotid bruit, and upper limb claudication due to arterial insufficiency. Its manifestations can be atypical, isolated and/or catastrophic disease [3]. Aortic aneurysm is the most common finding in such cases [5]. Here, we report our experience with a case of Takayasu`s Arteritis in a 42-year-old male who had an aortic root aneurism and underwent ascending aortic surgery.

Case report

Our patient was a 42-year-old male with a history of hypertension and a confirmed diagnosis of Takayasu arteritis, characterized by multiple arterial aneurysms affecting the left subclavian artery, both the left and right iliac arteries, and a distal abdominal aortic aneurysm. Ten years prior to this admission, he underwent an endovascular abdominal aorta repair. Since then, he has been attending outpatient follow-ups every six months, during which he had regular echocardiographic examinations and CT angiography.

Throughout his follow-up, the patient developed an ascending aortic aneurysm that progressively increased in size. It was monitored until it exceeded 5.5 cm in diameter, ultimately measuring 5.7 cm on transthoracic echocardiography. At this point, he was admitted for further evaluation and surgical intervention. Upon admission, the patient was asymptomatic,

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with all laboratory results within normal limits, and his echocardiography indicated normal cardiac function.

A cardiac CT scan revealed a tri-commissural aortic valve with significant aneurysmal dilation of the aortic root, particularly involving the right aortic sinus, which exhibited mild eccentric wall calcification (right aortic sinus dimensions: 41 x 36 x 41 mm; sinus of Valsalva diameter: 58 mm). The ascending aorta was of normal size (32 mm), with an unobstructed left aortic arch and isthmus. There was stable fusiform dilation of the tortuous bilateral subclavian arteries, left common carotid artery, and right dominant circulation. The right coronary artery originated from the aneurysmal right aortic sinus at an acute angle but showed no signs of calcified plaque, stenosis, or narrowing.

Following a discussion in the heart team meeting, the decision was made to proceed with surgical repair. In the operating room, the patient was positioned supine, and general anesthesia was administered. Central and femoral lines were inserted, and after the usual sterile preparation, a median sternotomy was performed. The pericardium was opened, and a pericardial well was created to facilitate dissection around the aneurysm. A full dose of heparin was administered, followed by aortic and venous cannulation.

Prolene 5-0 sutures were applied around the edge of the aneurysm, utilizing multiple plication sutures (four in total). We then reduced the flow and secured the sutures, maintaining hemostasis throughout the procedure. An equine patch was placed over the aneurysmal area, which was subsequently repaired and reinforced with BioGlue. The patient was weaned from cardiopulmonary bypass without complications. After removing the venous cannula and achieving hemostasis, protamine was administered, and the aortic cannula was removed, and hemostasis was performed.

The pericardium was reapproximated, and the sternum was secured with seven stainless steel wires. Finally, the subcutaneous tissue and fascia were closed with Vicryl sutures, while the skin edges were approximated using 4-0 Monocryl sutures. The patient tolerated the operation smoothly and was transferred to the cardiac surgery intensive care unit in stable condition.

Discussion

The Chapel Hill Consensus Conference defines TA as "granulomatous inflammation of the aorta and its major branches" within the context of systemic vasculitis nomenclature [6]. Due to the obstruction of large arteries arising from the aorta, it is also referred to as "pulseless disease" or "occlusive thromboaortopathy." Vascular changes can lead to various complications, including hypertension, often due to renal artery stenosis or stenosis of the suprarenal aorta; aortic valve involvement resulting in aortic insufficiency; arterial or aortic aneurysms; and pulmonary hypertension [3]. Renal arteries are involved in 24- 68% of cases, frequently bilaterally.

The exact pathophysiology remains unclear. The underlying mechanism involves inflammation, and several etiological factors have been identified. These include infection by Mycobacterium tuberculosis, spirochetes, and streptococcal species, as well as circulating antibodies linked to autoimmune processes. These factors contribute to the genetic susceptibility observed in some patients [3]. Pan-arteritis, characterized by significant intimal hyperplasia, adventitial and medial thickening, and infiltration of mononuclear and giant cells, is proposed as a primary etiology [4]. The disease is more prevalent in Asian women and typically peakes in their 30s.

Takayasu's arteritis is associated with substantial morbidity and can be life-threatening. It is a chronic, remitting, and relapsing disorder with an overall survival rate of 90%, which may decrease in the presence of complications. The disease can be classified into six types based on angiographic involvement [3], with our patient categorized as type II due to the presence of an ascending aortic aneurysm.

Management strategies depend on the severity of complications and the disease's activity, ranging from mild to severe deterioration.

Corticosteroids remain the cornerstone of treatment for active disease. In addition, interleukin-6 receptor inhibitors (tocilizumab) and B-cell depleting agents (rituximab) have shown clinical efficacy. Cytotoxic agents are reserved for relapsing or steroid-resistant cases, while anti-tumor necrosis factor agents have also demonstrated promising results in recurrent cases [7]. A recent case study reported a 26-year-old woman with Takayasu's arteritis presenting with thickened aortic arch walls and its branches. She experienced dizziness, lightheadedness, and syncope. The diagnosis was facilitated by the absence of radial and brachial pulses on the right side, along with a significant blood pressure difference between her arms. MRI revealed aberrant thickening of the aortic arch walls, measuring 4.4 mm in diameter. The right and left common carotid arteries, as well as the brachiocephalic and subclavian branches, exhibited wall thickening, confirming an active phase of Takayasu's arteritis. The therapeutic regimen included methotrexate, prednisolone, folic acid, and aspirin, with the patient showing clinical improvement by discharge and at follow-up [4].

Surgical revascularization is increasingly recognized as a primary treatment option [1]. During remission, critical stenotic lesions should be addressed through surgical revascularization or angioplasty. Bypass graft surgery offers the best long-term patency rates, while percutaneous balloon angioplasty can be effective for short lesions. Recurrent stenosis may be treated with stenting and angioplasty; however, conventional stents tend to have high failure rates in Takayasu's arteritis. Revascularization and aneurysm clipping are also viable options [7]. Indications for surgical therapy include coronary artery stenosis leading to myocardial ischemia, renal vascular stenosis causing hypertension, critical stenosis affecting three or more cerebral vessels and/or resulting in cerebral ischemia, and abdominal or aortic aneurysms with a diameter greater than 5 cm, as well as aortic regurgitation and severe aortic coarctation. Many patients may experience restenosis at the angioplasty site within 1-2 years after percutaneous transluminal coronary angioplasty [7]. Its overall role in treating Takayasu's arteritis is limited, as long-term benefits are often diminished by the fibrous nature of arterial obliteration [1].

Arterial reconstruction is another surgical option, tailored to the patient's anatomy and the location of the lesions. Major reconstructions of aortic lesions typically utilize polytetrafluoroethylene or Dacron, while autogenous saphenous veins are preferred for extremity or isolated mesenteric revascularization. Anastomoses are established on the relevant arterial segments to prevent early graft failure [1]. Ascending aortic aneurysms involving the proximal aortic arch, arising from any point between the aortic valve and the innominate artery, signal the need for open surgery. The Bentall procedure replaces the ascending aorta and aortic valve with a composite graft that re-implants the coronary arteries [8]. The David procedure offers a valve-sparing aortic root replacement, effectively treating aortic root aneurysms and improving long-term survival rates while minimizing complications [9]. Aortic root replacement combined with hemiarch replacement presents a low mortality risk and is indicated when the aortic root aneurysm extends to the distal ascending aorta [10]. Linear plication, along with external wrapping, represents another option supported by promising mid-term outcomes for patients with ascending aortic aneurysms at high surgical risk [10]. Most young patients with Takayasu's arteritis tolerate surgical therapy well. They often experience significant postoperative symptom relief postoperatively, including a reduction or complete resolution of hypertension after renovascular reconstruction and relief from upper limb ischemia following bypass surgery.

Conclusion

Takayasu's arteritis poses significant clinical challenges due to its potential for severe vascular complications. Our case illustrates that surgical intervention can be an effective management strategy, particularly in patients with type II Takayasu's arteritis who present with significant vascular anomalies, such as a large ascending aortic aneurysm. The successful surgical repair of

our patient underscores the importance of timely intervention.

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References

1. Trinidad B, Surmachevska N, Lala V. "Takayasu Arteritis." StatPearls, StatPearls Publishing, 8 August 2023.
2. Tombetti E, Mason JC. Takayasu arteritis: advanced understanding is leading to new horizons." Rheumatology (Oxford, England). 2019; 58(2): 206-219.
3. Arafat, A.A. Surgery for autoimmune aortitis: unanswered questions. Cardiothorac Surg. 2019; 27 (4): 1-6.
4. Khadka A, Singh S, Timilsina S. Takayasu's Arteritis: A Case Report. JNMA; journal of the Nepal Medical Association. 2022; 60 (56): 1041-1044.
5. Svensson LG, Arafat A, Roselli EE, et al. Inflammatory disease of the aorta: patterns and classification of giant cell aortitis, Takayasu arteritis, and nonsyndromic aortitis. The Journal of thoracic and cardiovascular surgery; 2015; 149 (2 Suppl): S170-5.
6. Jennette JC, Falk RJ, Andrassy K, et al. Nomenclature of systemic vasculitides. Arthritis and rheumatism. 1994; 37 (2): 187-92.
7. Clifford AH, Arafat A, Idrees JJ, et al. Outcomes Among 196 Patients With Noninfectious Proximal Aortitis. Arthritis & rheumatology (Hoboken, N.J.). 2019; 71 (12): 2112-2120.
8. Sran S, Sran M, Ferguson N, Makaryus AN. Surgical management of ascending aortic aneurysm and its complications. Case reports in vascular medicine. 2014: 102605.
9. Espitia O, Bruneval P, Assaraf M, et al. Long-Term Outcome and Prognosis of Noninfectious Thoracic Aortitis. Journal of the American College of Cardiology. 2023; 82 (11): 1053-1064.
10. Malaisrie SC, Duncan BF, Mehta CK, et al. The addition of hemiarch replacement to aortic root surgery does not affect safety. The Journal of Thoracic and Cardiovascular Surgery. 2015; 150(1) 118-124.