CASE REPORT

Idiopathic subvalvular aortic aneurysm masquerading as acute coronary syndrome

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SUMMARY

Subvalvular aneurysms are the least common type of left ventricular (LV) aneurysms and can be fatal. Subaortic LV aneurysms are much rarer than submitral LV aneurysms and mostly reported in infancy. They can be congenital or acquired secondary to infections, cardiac surgery or trauma. Here, we report a unique presentation of a large, idiopathic subaortic aneurysm in an adult masquerading as an acute coronary syndrome. Diagnosis was made with the help of a CT aortography. Aneurysm was surgically resected with good results. This case highlights the clinical presentation and management of subaortic aneurysms, an important differential for congenital aortic malformations.

BACKGROUND

Subvalvular aneurysms are rare, relatively unknown cause of sudden death in young individuals. They can be submitral or subaortic in location based on the relationship of the orifice of the aneurysm to the annulus of the affected valve.^{1 2} This condition was originally described in people of African ancestry and increasingly now in other races as well.^{3–5} The congenital forms are postulated to arise as a result of a developmental weakness below the level of the mitral or aortic valve due to defective fusion of the myocardium with the valve annulus.^{1 6 7} However, most cases arise as a sequelae of myocardial infarction,⁸ myocarditis due to tuberculosis, rheumatic fever, Chagas' disease, syphilis and bacterial endocarditis.^{9–11} In the initial stages, they are mostly asymptomatic, and patients typically present in the second or third decade of life with heart failure from weakening of the annular ring and valvular incompetence.³ Life-threatening complications result from sudden rupture or extrinsic compression of nearby structures causing myocardial infarction (MI), malignant arrhythmias and thromboembolism.⁷

CASE PRESENTATION

A 39-year-old Asian man presented with sudden-onset substernal chest pain radiating to the back, shortness of breath, diaphoresis and vomiting. He has also been experiencing gradually worsening exercise intolerance over the past 2 years. He does not have a past history of syphilis, rheumatic fever, tuberculosis or bacterial endocarditis. He denies any chest trauma, thoracic surgeries or prior heart disease. He smokes 1 pack of cigarettes every week since 10 years and occasionally consumes alcohol. He denies any intravenous drug use. Family history

is negative for heart disease or sudden cardiac death. Physical examination revealed a heart rate of 104/min in sinus rhythm, a blood pressure of 80/50 mm Hg and a 2/6 systolic murmur in the right second intercostal space without radiation. Peripheral pulses and blood pressures were bilaterally equal in both upper extremities.

INVESTIGATIONS

ECG showed diffuse T inversions in anterolateral chest leads with right ventricular hypertrophy pattern. Serial troponin levels were elevated. Chest X-ray showed cardiomegaly and mediastinal widening (figure 1). 2D Doppler echocardiography showed a wedge-shaped echo free space between ascending aorta and left atrium with mild aortic regurgitation (figure 2). A CT aortography was obtained, which strikingly revealed a trilobed annular subaortic aneurysm in the atrioventricular groove measuring 9.6 cm transversely, 8.6 cm craniocaudally and 6.4 cm anteroposteriorly. It was communicating with the left ventricular outflow tract (LVOT). Another lobulated aneurysmal sac was seen anterior to the right coronary cusp, which

Figure 1 Chest X-ray taken at presentation showing cardiomegaly and widened mediastinum.

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Figure 2 2D echo colour flow image showing three flows: (1) aortic regurgitation due to distortion of aortic valve by the aneurysm; (2) large flow into Aneurysm 1 between left atrium and ascending aorta; (3) smaller jet into Aneurysm 2.

appeared to communicate with the right coronary sinus and dissect into the interventricular septum (figure 3A, B). Selective trans-radial coronary angiography revealed extrinsic compression of the proximal left circumflex and right coronary arteries by the aneurysms; however, no calcification or atheromatous plaque was visualised (figure 4A, B).

DIFFERENTIAL DIAGNOSIS

Aortic dissection or rupture and Boerhaave's syndrome (oesophageal rupture) are other common causes for acute chest pain with mediastinal widening on chest X-ray. Differentials for aortic aneurysms include other conotruncal congenital or acquired defects such as sinus of Valsalva (SOV) aneurysm, aortico-left ventricular (LV) tunnel, and aortic root and LV pseudoaneurysms.

TREATMENT

Surgical repair of the aneurysm was attempted through a median sternotomy approach and cardiopulmonary bypass. Intraoperatively, a trilobed aneurysm was identified (figure 5A). There were two openings beneath the aortic leaflets in the LVOT, feeding the aneurysm sacs. The larger of the two

Figure 4 (A) Coronary angiography image showing external compression of the RCA by Aneurysm 1. (B) External compression of the LCA by Aneurysm 2. LCA, left coronary artery; RCA, right coronary artery.

openings was found just inferior to non-coronary cusp and was repaired with a polytetrafluoroethylene patch. The second mouth just inferior to the left coronary cusp was directly closed with 4/0 polypropylene suture (figure 5B). Specimen of the wall of aneurysm was resected and sent for histopathology. The aortic valve leaflets, ascending aorta and all three aortic sinuses were of normal morphology (figure 5C).

OUTCOME AND FOLLOW-UP

The patient had an uneventful hospital course following surgery and was found to be in NYHA Class 1 at 1-month and 6-month follow-up visits postsurgery.

DISCUSSION

Subaortic annular aneurysms are less common than submitral aneurysms and mostly evidenced in the literature as isolated case reports. Hence, the true incidence of this condition in the general population is still largely unknown. They are difficult to diagnose because of lack of characteristic findings on routine imaging modalities like chest X-ray and 2D echocardiogram. However, with improvement in imaging techniques and wider availability, these aneurysms are recognised and reported more frequently than before. Previous case reports have described coronary arterial obstruction from external compression by large submitral LV aneurysms, but not so often with subaortic LV aneurysms.^{3 12} This is because subaortic aneurysms mostly

Figure 3 CT aortography image showing a large trilobed subaortic aneurysm (1) in relation to the non-coronary cusp and a smaller subaortic aneurysm (2) in relation to the left cusp of the aortic valve.

Figure 5 (A) Intraoperative appearance of the trilobed subaortic aneurysm after releasing pericardial adhesions. (B) The mouth of a smaller second aneurysm opening into the aorta below the left cusp of the aortic valve, closed with sutures. (C) Normal-appearing tricuspid aortic valve.

Figure 6 Histology slide of the aneurysm wall showing (A) the absence of elastic fibres on Verhoeff's stain and (B) showing blue staining of collagen fibres with Masson trichome stain.

occur in relation to the left coronary cusp of the aortic valve, are more restricted anatomically and do not reach large sizes as in the case of submitral aneurysms.¹ Interestingly, in our patient, the aneurysm was much larger when compared to the aortic diameter and demonstrated severe compression of the coronary arteries on angiogram.

Subannular aneurysms are in reality 'false' aneurysms as they are devoid of all structural elements of a regular aortic wall. Most reported cases are acquired secondary to infections and post-MI; however, in our patient, there was no apparent aetiology for the development of the subaortic aneurysm. Tuberculosis and syphilis serology were negative. The histology of the resected specimen showed fibrocollagenous tissue with hyalinisation and the absence of cardiac muscle or aortic media representing a 'false' aneurysm morphology and additionally showed no evidence of inflammation or infection (figure 6A, B). Subaortic aneurysms can co-occur with other conotruncal anomalies like an SOV aneurysm or other congenital anomalies like bicuspid aortic valve, atrial or ventricular septal defects and coarctation of aorta.^{6 10 13} Our patient did not have any such coexistent pathology, and the aortic valve was trileaflet with normal morphology as confirmed intraoperatively (figure 5C).

Transoesophageal echocardiogram is superior to transthoracic echocardiogram in identifying the location of the neck of the aneurysm and in delineating the extent of the aneurysm.¹⁴ However, non-invasive modalities like CT angiography and cardiac MRI are now recognised as standard tests to diagnose and characterise subvalvular aneurysms as they also aid in surgical planning.^{8 15} Small asymptomatic aneurysms can be managed conservatively with frequent imaging surveillance. Surgical resection of aneurysm is the treatment of choice in all haemodynamically unstable patients and those who present with compressive symptoms.⁷ Surgical options include the traditional Bentall procedure that involves replacement of the aortic root

with a composite graft and an implanted mechanical valve. This also involved reimplantation of the coronary arteries into the graft. Various modifications to this procedure including remodelling or reimplantation techniques have also been carried out with varying results.¹⁶ Increasingly, valve-sparing aortic root procedures have been preferred by surgeons over the Bentall surgery for two major reasons: first, lifelong anticoagulation and associated bleeding complications can be avoided. Patients report a better overall quality of life following valve-sparing surgeries when compared to the Bentall procedure, especially in young individuals.¹⁷ More importantly, a recent meta-analysis by Arabkhani *et al*¹⁸ showed low pooled early and late mortality

Learning points

- ▶ Suspect a subvalvular aortic aneurysm in any young, non-hypertensive adult presenting with mediastinal widening on chest X-ray with clinical symptoms concerning for an acute coronary syndrome.
- ▶ Subvalvular aneurysms can cause death from sudden rupture or from external compression of nearby structures, especially the coronary arteries if not diagnosed early.
- ▶ Search for secondary causes like tuberculosis, rheumatic fever, syphilis and bacterial endocarditis in all cases of subvalvular aneurysms.
- ▶ Investigate for other conotruncal anomalies like aortic coarctation, bicuspid aortic valve and atrial/ventricular septal defects in all diagnosed cases.
- ▶ Early surgical approach is the definitive management of all symptomatic, large subvalvular aortic aneurysms.

rates, and lower incidence of haemorrhage, thromboembolism and infective endocarditis following valve-sparing surgeries.

This brief report aims to educate the readers regarding the various aetiologies, natural history, complications and management of subaortic annular aneurysms. In the absence of standardised treatment options, decision to operate and the type of surgery should be individualised based on patient preferences. They should be educated regarding potential consequences of an unrepaired aneurysm. If conservative management is chosen, then they should be closely followed up with frequent imaging. Long-term follow-up of surgically corrected aneurysm is also important as recurrence following surgery is possible but has not been reported that often.

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