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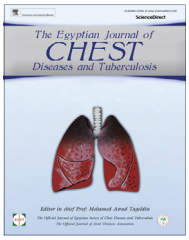

CASE REPORT

Dissecting aortic root aneurysm and severe aortic regurgitation following pulmonary tuberculosis

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KEYWORDS

Aneurysm;
Aortitis;
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Aortic regurgitation;
Aortic root

Abstract Aneurysms of the aortic root and ascending aorta are often due to degenerative disease of media but tuberculosis is an important but extremely rare cause of aortic root dilatation especially in tropical countries like India where tuberculosis is endemic. Tubercular aneurysmal dilatation of aorta with dissection leading to aortic regurgitation is a rare but important complication of tuberculosis. With worldwide resurgence of tuberculosis due to increasing incidence of drug-resistant tuberculosis and its association with acquired immunodeficiency syndrome, the tubercular aneurysm has become a real clinical entity. Although tubercular aortitis is fairly common, tuberculous mycotic aneurysm of the aorta is rare, with involvement of the aortic root being exceedingly uncommon.

Here we describe a case of 18 year old male presenting with severe breathlessness and was found to have dissecting aortic root aneurysm with aortic regurgitation with active pulmonary tuberculosis and spinal deformity in the form of kyphoscoliosis. He underwent a Bentall procedure, and excised aortic root tissue showed epithelioid cell granulomas with panarteritis.

Case report

A 18-year-old male was admitted with complaints of recurrent chest pain, progressive dyspnea with exertion, and inability to walk for more than 100 m. He complained of fatigue, weight loss, night sweats, and adenopathy of his left axilla and neck for past 3 months. Physical examination revealed diastolic murmur in right second intercostal space with visible pulsation in the suprasternal region and neck, apex beat was displaced downwards into the 6th intercostal space. His admission chest radiograph demonstrated cardiomegaly, left sided lung was markedly destroyed due to active pulmonary tuberculosis,

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chest radiograph also suggestive of marked spinal deformity in the form of kyphoscoliosis probably due to extensive pulmonary fibrosis or as a sequel of tubercular infection of spine. The patient underwent cardiac evaluation with 2-dimensional echocardiography which revealed a severely dilated aortic root measuring about 55 mm with severe aortic regurgitation, left ventricular ejection fraction of 55%. Patient underwent cardiac computed tomography which demonstrated dissection and dilation of aortic root and ascending aorta rest of the aorta is normal in caliber also the coronary arteries were normal. Patient was a known case of primary pulmonary tuberculosis for which he received antitubercular 4-medication regimen for 3 months.

After his full workup patient and attendants were fully explained about the whole scenario and clinical condition and about the high risk involved in surgery. After taking informed consent the patient was operated and Bentall procedure was done with replacement of incompetent aortic valve and dilated ascending aorta with valved conduit of 25 mm mechanical valve size. The excised aortic tissue was sent for histopathological examination which revealed panarteritis with epithelioid cell granulomas in background of chronic inflammatory cells.

In postoperative period the patient was ventilated for 36 h and was then extubated. In immediate postoperative period the patient maintained his hemodynamic status and was on minimal inotropic support which was soon weaned off and also patient drained only about 300 ml of blood through chest and mediastinal drains which was removed on the second postoperative day and the patient was started treatment with injectable streptomycin. Patient maintained normal blood gases for almost 48 h after that patient stated developing tachypnoea and started retaining carbonmonoxide in blood and became drowsy following this the patient was immediately reventillated electively, the patient improved and was again extubated and aggressive chest physiotherapy was started but the patient was not able to maintain normal blood gases and was again ventilated. Apart from respiratory dysfunction patient was maintaining his hemodynamic status very well and without any inotropic supports, his renal and hepatic functions were also within normal range. Later the patient developed acute respiratory distress syndrome as evident by fluffy opacities found in his chest X ray. Despite the aggressive antibiotic cover and ventilatory support patient succumb to his illness after 10 days of surgery due to respiratory failure (Figs. 1–5).

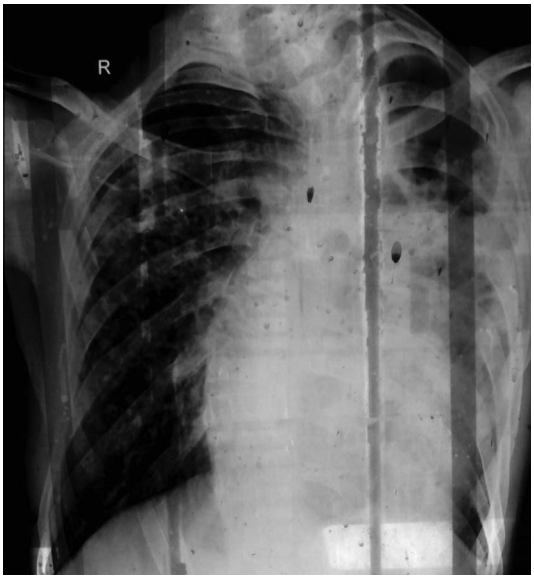

Figure 1 X ray chest showing destroyed left lung with cardiomegaly with spinal deformity in the form of kyphoscoliosis.

Figure 2 Angiographic film showing dilated aortic root with dissection.

Discussion

Dissecting aortic aneurysms is most often caused by atherosclerosis, other causes includes:

- Marfan syndrome (a genetic connective tissue disorder)
- Other non-specific connective tissue disorders (characterized by a family history of aneurysms)
- Presence of a bicuspid aortic valve
- Syphilis
- Tuberculosis and trauma are rare causes

Tuberculous dissecting aneurysms of the aorta is exceedingly rare, but the lesion is uniformly fatal if not diagnosed promptly. Typical clinical scenarios include evidence of tuberculous lymph nodes in 70% of cases, with 1 or more of 3 presentations: (1) fever and persistent pain related to the location of the aneurysm, (2) hypovolemic shock or other evidence of massive bleeding, or (3) pulsatile, rapidly expanding paraaortic mass [1]. The perforation of the aortic wall is generally surrounded by thrombotic debris and inflammatory tissue [2]. Out of these presentations our patient has enlarged cervical lymph nodes with complains of fever and weight loss and dyspnoea and chest pain.

Establishing the diagnosis of infectious aortitis or mycotic aortic aneurysm early is critical because this condition is associated with a high rate of rupture and subsequent mortality if left untreated [9–13]. The diagnosis of tuberculous aortitis is very difficult to establish because this disorder is exceedingly rare and can mimic Takayasu arteritis [14]. Delay in diagnosis

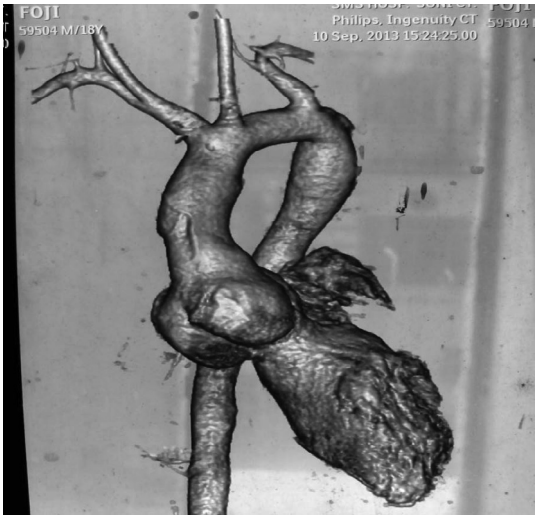

Figure 3 Angiographic film showing dilated aortic root with dissection.

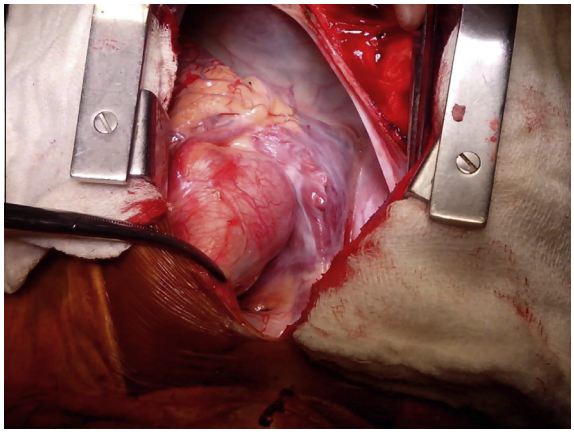

Figure 4 Intraoperative photograph showing dilated aortic root.

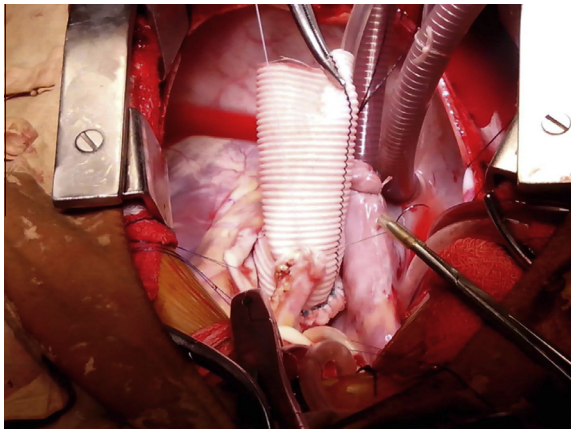

Figure 5 Intraoperative photograph showing dilated portion of aortic root removed and replaced by valved conduit and coronary buttons anastomosed to valved conduit.

may occur, and patients may be treated with immunosuppressive therapy before the diagnosis of tuberculosis is made. Thus, the possibility of tuberculous aortitis should be considered among patients with aortitis or atypical aortic aneurysms that have a history of pulmonary or extrapulmonary tuberculosis or chronic immunosuppression or who present with a suspicious finding such as a cavitary lung lesion, pleural effusion, or lymphadenitis [14]. In such cases, a definitive diagnosis should be pursued before glucocorticoids are given.

Most patients with dissecting aortic aneurysms are asymptomatic at the time of diagnosis, because the aneurysms are typically discovered incidentally on imaging studies (chest X-ray, CT scan, or echocardiogram) ordered for other indications. Aneurysms of the root or ascending aorta may produce secondary aortic regurgitation, so a diastolic murmur may be detected on physical examination or, less often, patients may present with congestive heart failure. When dissecting aortic aneurysms are large, patients may suffer a local mass effect, such as compression of the trachea or mainstem bronchus (causing cough, dyspnea, wheezing, or recurrent pneumonitis), compression of the esophagus (causing dysphagia), or compression of the recurrent laryngeal nerve (causing hoarseness). Rarely, chest or back pain may occur with nondissecting aneurysms as a result of direct compression of other intrathoracic structures or erosion into adjacent bone.

The feared consequence of thoracic aneurysms is aortic dissection or rupture (often referred to as an acute aortic syndrome), which is potentially lethal. Typical symptoms of acute aortic syndrome include the abrupt onset of severe pain in the chest, neck, back, and/or abdomen.

Often, dissecting aortic aneurysms is evident on chest X-ray films and are characterized by widening of the mediastinal silhouette, enlargement of the aortic knob, or tracheal deviation. However, smaller aneurysms and even some large ones may not produce any abnormalities on chest X-ray films. One should be cautious in interpreting a radiology report that describes an abnormal aortic silhouette as an ''ectatic aorta''. If a chest X-ray film shows an enlarged aortic silhouette, one should have a low threshold for ordering a tomographic imaging study (e.g., CT scan) to define the aortic anatomy.

Contrast-enhanced CT scanning and MR angiography are the preferred modalities to define aortic (and branch vessel) anatomy, and both accurately detect and size thoracic aortic aneurysms.

Transthoracic echocardiography is effective for imaging the aortic root and is thus often used to evaluate patients with Marfan syndrome. However, it does not consistently visualize the mid or distal ascending aorta well, nor does it well visualize the descending aorta. Therefore, other than those with Marfan syndrome, transthoracic echocardiography should generally not be used for diagnosing and sizing thoracic aortic aneurysms. Although transesophageal echocardiography can visualize the entire thoracic aorta well, given its semi-invasive nature it is not favored for the routine imaging of those with stable (nondissecting) thoracic aneurysms.

On the basis of longitudinal data from the Yale group, Davies et al. [3] found that the mean rate of growth for all thoracic aortic aneurysms was 0.1 cm/y. The rate of growth, however, was greater for aneurysms of the descending aorta versus ascending aorta, was greater for dissected aneurysms versus nondissected ones, and was greater for those with Marfan syndrome versus those without. Initial size is an important

predictor of the rate of aortic aneurysm growth. However, even controlling for initial aneurysm size, there is still substantial variation in individual aneurysm growth rates [4], thus making it difficult to prospectively predict growth for a given aneurysm. Therefore, all aneurysms need to be followed up with regular surveillance imaging to monitor growth. With regard to aneurysm size and the risk of rupture or dissection, in their series Davies et al. [3] found an annual rate of 2% for aneurysms <5 cm, 3% for aneurysms 5–5.9 cm, and 7% for aneurysms $\geqslant 6$ cm in diameter. Therefore, the risk appears to rise abruptly as aortic aneurysms reach a size of 6 cm.

The optimal timing of surgical repair of aortic aneurysms remains somewhat uncertain, given the limited data on their natural history. For most ascending thoracic aortic aneurysms, surgery is indicated at a diameter of $\geqslant 5.5$ cm. Among those with an increased operative risk (e.g., the elderly or those with comorbidities), it is typically advised to raise the threshold to 6 cm or more before recommending surgery. Conversely, among patients who are at increased risk of aortic dissection or rupture (e.g., Marfan syndrome or bicuspid aortic valve), it is often recommended to perform an ascending aortic repair when aneurysms reach only 5 cm and in selected cases (those at especially high risk), at even smaller diameters [5]. Moreover, when patients with a bicuspid valve require aortic valve replacement surgery, it is recommended to perform a prophylactic replacement of the ascending aorta if its diameter is 4 cm or greater, given that we now recognize that such patients would otherwise remain at high risk for subsequent aortic dissection. For most descending thoracic aortic aneurysms, surgery is recommended at an aortic diameter of 6 cm or greater. The mortality of elective surgical repair of ascending aortic aneurysms in large centers is 3–5%. Aortic repair requires cardiopulmonary bypass, and the aneurysm is generally resected and replaced with a prosthetic tube graft of appropriate size. When the aneurysm involves the aortic root and is associated with significant aortic regurgitation, one usually performs a composite aortic repair Bentall procedure.

Alternatively, when the aortic valve leaflets are structurally normal a valve-sparing root replacement may be performed. This technique, advanced by Dr. Tirone David, involves excising the sinuses of Valsalva while sparing the aortic leaflets, sewing a Dacron graft to the base of the aortic annulus, and reimplanting the aortic valve leaflets within the graft to restore their normal anatomic configuration [6]. When successful, this procedure avoids the need for prosthetic valve replacement and the associated long-term risks and reduces the risk for repeated valve surgery. In a series of 151 patients with aortic root aneurysms who underwent valve-sparing surgery, David et al. [6]found that at 8 years of follow-up, 67% had mild or no aortic regurgitation, whereas only 2% developed severe aortic regurgitation.

Some younger patients have a dilated aortic root, but because of intrinsic valve dysfunction, the aortic valve cannot be spared. For those wishing to avoid the prosthetic valve required with the composite aortic graft, one option has been a pulmonary autograft, also known as the Ross procedure. In this procedure, the patient's aortic valve and root are excised. The patient's own pulmonary root is then excised and transplanted into the aortic position. In one series of 91 patients, the incidence of pulmonary autograft root dilatation was 61% at 5 years among those with a prior aortic aneurysm, compared with 27% among those with no prior aortic aneurysm (i.e., those who had surgery for aortic valve disease only) [7].

When full aortic root/valve replacement is necessary, another alternative to a composite graft is the use of cryopreserved aortic allografts (cadaveric aortic root and proximal ascending aorta). However, after the procedure, late structural valvular deterioration does occur and may lead to subsequent reoperation. Moreover, the aortic valve deterioration appears to progress more rapidly in younger patients than in older ones, making the homograft less likely to serve as a durable, lifelong prosthesis [8].

In the present case we excised the aortic leaflets and aneurysmal aorta and replaced it with valved conduit composite graft on cardiopulmonary bypass. In a country like India where tuberculosis is still endemic tuberculosis is an important but extremely rare cause of mycotic aortic aneurysm.

Conclusion

Aneurysm of the aortic root often affects patients in their second to fourth decades of life, whereas aneurysm of the ascending aorta occurs mostly in the fifth to seventh decades of life. These aneurysms can cause aortic insufficiency, dissection, and/or rupture. Current guidelines recommend surgical treatment when the diameter of the aneurysm exceeds 50 mm. In patients with family history of aortic dissection or with Loyes–Dietz syndrome (a more severe form of Marfan syndrome), surgery should be considered when they are even smaller. Composite replacement of the aortic valve and ascending aorta used to be the standard treatment for patients with aortic root aneurysms. Because aneurysms are often first detected on an imaging study ordered for other indications, any suggestion of an enlarged aorta should prompt follow-up with an appropriate dedicated imaging study. Fortunately, modern imaging techniques—especially CT and MRI—have now made the sizing and surveillance of aneurysms relatively easy. In the future, genetic screening may also play a role in the screening of those with a family history of thoracic aortic aneurysms. Combined antitubercular chemotherapy and surgical intervention seem to offer the best chance for a cure and constitute the only way to salvage patients with this disease.

References

[1] R. Long, R. Guzman, H. Greenberg, J. Safneck, E. Hershfield, Tuberculous mycotic aneurysm of the aorta: review of published medical and surgical experience, Chest 115 (1999) 522–531.
[2] S.K. Choudhary, A. Bhan, S. Talwar, M. Goyal, S. Sharma, P. Venugopal, Tubercular pseudoaneurysms of aorta, Ann. Thorac. Surg. 72 (2001) 1239–1244.
[3] R.R. Davies, L.J. Goldstein, M.A. Coady, S.L. Tittle, J.A. Rizzo, G.S. Kopf, J.A. Elefteriades, Yearly rupture or dissection rates for thoracic aortic aneurysms: simple prediction based on size, Ann. Thorac. Surg. 73 (2002) 17–28.
[4] O.E. Dapunt, J.D. Galla, A.M. Sadeghi, S.L. Lansman, C.K. Mezrow, R.A. de Asla, C. Quintana, S. Wallenstein, A.M. Ergin, R.B. Griepp, The natural history of thoracic aortic aneurysms, J. Thorac. Cardiovasc. Surg. 107 (1994) 1323–1332.
[5] R.B. Devereux, M.J. Roman, Aortic disease in Marfan's syndrome, N. Engl. J. Med. 340 (1999) 1358–1359.
[6] T.E. David, J. Ivanov, S. Armstrong, C.M. Feindel, G.D. Webb, Aortic valve-sparing operations in patients with aneurysms of

the aortic root or ascending aorta, Ann. Thorac. Surg. (2002) S1758–S1761.
[7] G.B. Luciani, G. Casali, A. Favaro, M.A. Prioli, L. Barozzi, F. Santini, A. Mazzucco, Fate of the aortic root late after Ross operation, Circulation suppl II (2) (2003), II-61–II-67.
[8] J.J.M. Takkenberg, M.J. Eijkemans, L.A. van Herwerden, E.W. Steyerberg, M.M. Lane, R.C. Elkins, J.D. Habbema, A.J. Bogers, Prognosis after aortic root replacement with cryopreserved allografts in adults, Ann. Thorac. Surg. 75 (2003) 482–1489.
[9] E.A. Foote, R.G. Postier, R.A. Greenfield, M.S. Bronze, Infectious aortitis, Curr. Treat. Options Cardiovasc. Med. 7 (2005) 89–97.
[10] J.A. Bardin, G.M. Collins, J.B. Devin, N.A. Halasz, Nonaneurysmal suppurative aortitis, Arch. Surg. 116 (1981) 954–956.
[11] M.L. Fernandez Guerrero, J.M. Aguado, A. Arribas, C. Lumbreras, M. de Gorgolas, The spectrum of cardiovascular infections due to Salmonella enterica: a review of clinical features and factors determining outcome, Medicine (Baltimore) 83 (2004) 123–138.
[12] B.T. Muller, O.R. Wegener, K. Grabitz, M. Pillny, L. Thomas, W. Sandmann, Mycotic aneurysms of the thoracic and abdominal aorta and iliac arteries: experience with anatomic and extra-anatomic repair in 33 cases, J. Vasc. Surg. 33 (2001) 106–113.
[13] D.J. Reddy, A.D. Shepard, J.R. Evans, D.J. Wright, R.F. Smith, C.B. Ernst, Management of infected aortoiliac aneurysms, Arch. Surg. 126 (1991) 873–878.
[14] A. Gajaraj, S. Victor, Tuberculous aortoarteritis, Clin. Radiol. 32 (1981) 461–466.