

Syphilitic Aortic Arch Aneurysm: Review of Cases

Somchai Waikittipong, MD

Department of Surgery, Yala Hospital, Yala, Thailand

Abstract

Syphilitic aortic aneurysms are uncommon today. Four cases of syphilitic aortic arch aneurysm are reported. The first case presented with upper airway obstruction. The second case presented with chest pain. The third case presented with dysphagia. And the last case presented with hoarseness. Three cases were successfully treated with aortic arch replacement. Pathophysiology and clinical manifestations of syphilitic aortic aneurysm are also reviewed.

Keywords: Syphilis, Aortic arch aneurysm

INTRODUCTION

Syphilitic aortic aneurysms are rarely seen today. These aneurysms can reach large proportions with compression and erosion into surrounding structures. Aneurysms of the transverse arch may impinge on the trachea, esophagus, or recurrent laryngeal nerve, resulting in stridor, dyspnea, dysphagia, hoarseness and pain. We review four cases of syphilitic aortic arch aneurysm, three of which were successfully treated with aortic arch replacement.

PATIENTS AND METHODS

From June 2011 to August 2014, there were four cases of syphilitic aortic arch aneurysm. The details of each case are presented below.

First Case

A 42 year-old man was referred to our hospital in June 2011 with symptoms of upper airway obstruction. Chest X-ray showed a large superior mediastinal mass.

On computed tomography scan, the mass was recognized as a saccular aortic arch aneurysm and measured 7.0 cm in maximal diameter. The trachea was compressed, narrowed, and displaced to the right (Figure 1). Serologic test for syphilis, both VDRL and TPHA, were positive. Computed tomography angio-

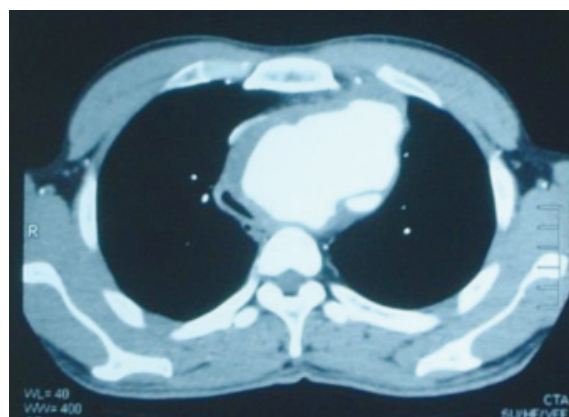


Figure 1 Computed tomography scan showing a large saccular aortic arch aneurysm. The trachea was compressed, narrowed, and displaced to the right.

Correspondence address: Somchai Waikittipong, Department of Surgery, Yala Hospital, Muang, Yala 95000, Thailand; Telephone: +668 9656 4408; Fax: +662 7321 2764; Email : somchaiwaikittipong@hotmail.com

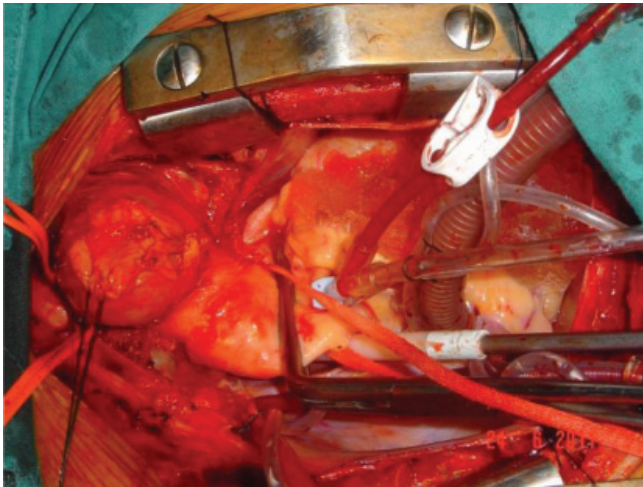


Figure 2 A large saccular aneurysm at the transverse arch.

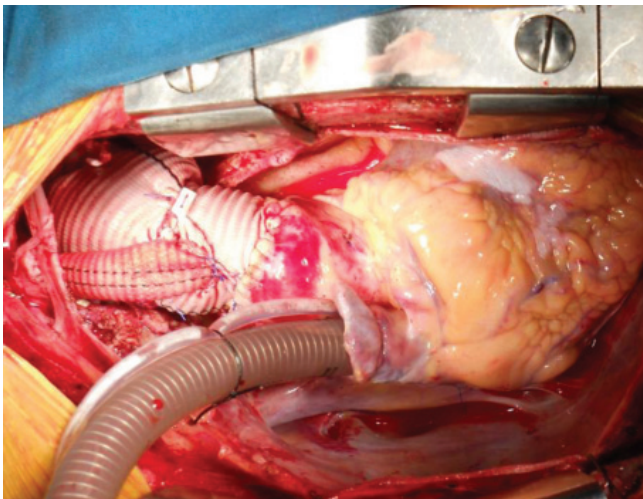


Figure 3 Partial aortic arch replacement with woven Dacron graft.

gram of the thoracic aorta demonstrated that the aneurysm originated from origin of the innominate artery to just before the origin of the left common carotid artery. Partial aortic arch replacement with woven Dacron graft was performed. The distal anastomosis was placed just proximal to the origin of left common carotid artery and the proximal anastomosis was placed at the ascending aorta, then a separate 10 mm graft was interposed from the origin of innominate artery to the side of the graft (Figures 2 to 3). The patient recovered well and was discharged home uneventfully. The pathological examination of resected aortic wall revealed inflammatory cell infiltration with plasma cell predominance (Figure 4).

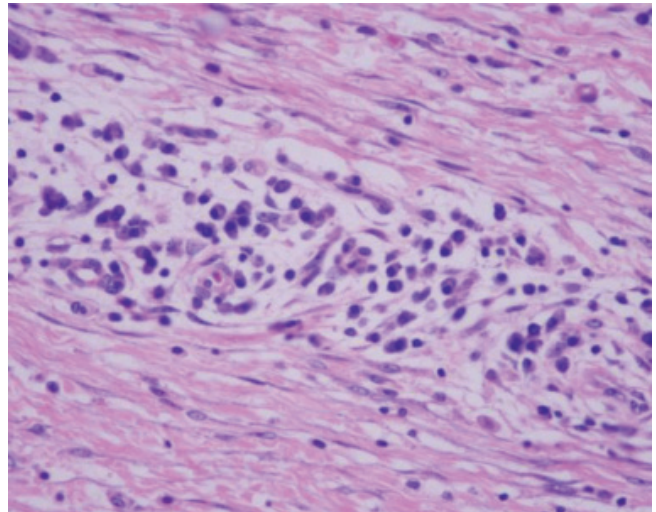


Figure 4 Pathological examination of resected aortic wall revealed inflammatory cell infiltration with plasma cell predominance.

Second Case

A 68 year-old man presented with chronic chest pain. Chest X-ray showed large a superior mediastinal mass. On computed tomography scan, a large saccular aneurysm of aortic arch was recognized which eroded into the chest wall (Figure 5). Serologic test for syphilis, both VDRL and TPHA were positive. Partial aortic arch replacement with woven Dacron graft was performed. The innominate artery and left common carotid artery were re-implanted with separate woven Dacron grafts. The patient recovered well. Pathological examination of the resected aortic wall was consistent with syphilitic aneurysm.

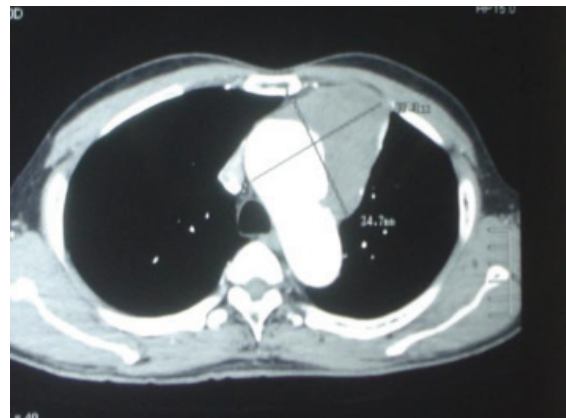


Figure 5 Computed tomography scan showed a large saccular aortic arch aneurysm which eroded into the chest wall.

Third Case

A 74 year-old woman presented with dysphagia. Chest X-ray and computed tomography scan revealed a large saccular aortic arch aneurysm. The aneurysm compressed the esophagus (Figure 6). Serologic test for syphilis, both VDRL and TPHA, were positive. The patient refused operation.

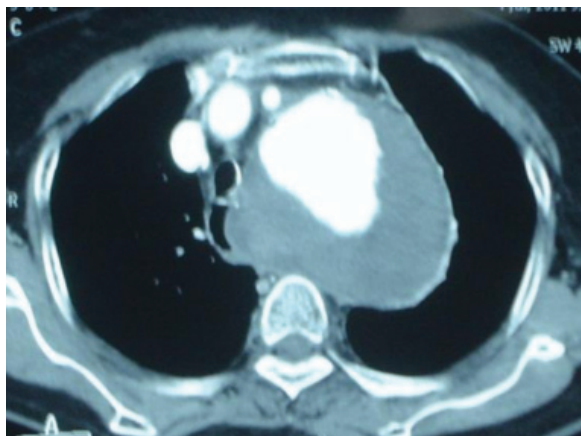


Figure 6 Computed tomography scan revealed a large saccular aortic arch aneurysm compressing the esophagus.

Forth case

A 61 year-old man presented with hoarseness. Chest X-ray and computed tomography scan revealed a large saccular aneurysm at the distal aortic arch (Figure 7). Serologic test for syphilis, both VDRL and TPHA, were positive. Total aortic arch replacement with four-branch woven Dacron graft was performed

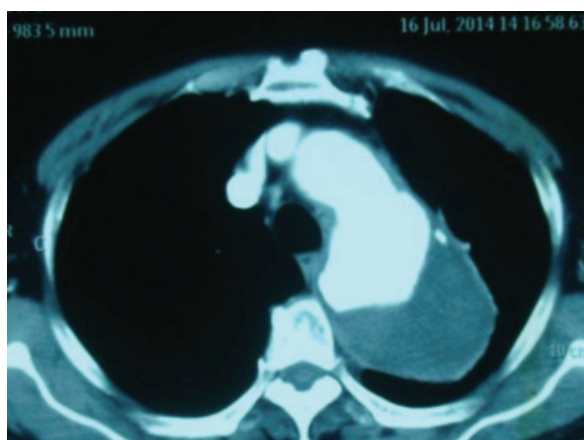


Figure 7 Computed tomography scan revealed a large saccular aneurysm at distal aortic arch.

uneventfully (Figure 8). Pathological examination of the resected aortic wall was consistent with syphilitic aneurysm (Figure 9).



Figure 8 Total aortic arch replacement with four-branch woven Dacron graft.

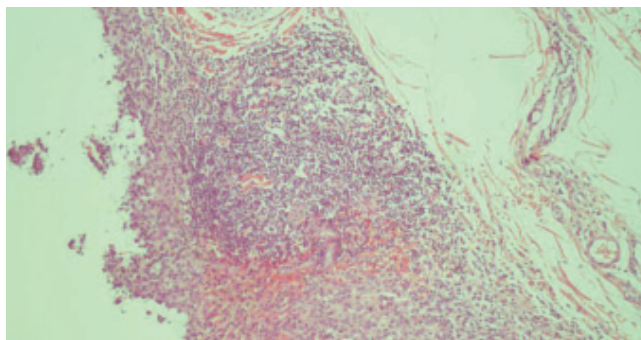


Figure 9 Pathological examination of resected aortic wall showing mixed, chronic, inflammatory cell (lymphocytes, plasma cells and histiocytes) infiltrate around the vasa vasorum with endarteritis obliterans

DISCUSSION

Syphilis was once a major cause of thoracic aortic aneurysms¹. Since the advancement of antibiotics, however, cardiovascular complications of syphilis are rarely seen today². Syphilis is a sexually transmitted infection caused by the spirochete bacteria *treponema pallidum* subspecies *pallidum*. The primary route of transmission is through sexual contact. Syphilis can present in one of four different stages: primary, secondary, latent, and tertiary. Without treatment a third of infected people develop tertiary disease³.

Tertiary syphilis may occur approximately 3 to 15 years after the initial infection and may be divided into three different forms: gummatous syphilis, late neurosyphilis, and cardiovascular syphilis. The basic lesion of cardiovascular syphilis is aortitis. Involvement of the cardiovascular system presumably begins early in the course of infection when the treponemes disseminate hematogenously from the site of inoculation. The organisms lodge within the nutrient blood vessels of the aortic adventitia and initiate the inflammatory changes.

The histologic changes begin as perivascular plasma cell and lymphocytic infiltrates of the vasa vasorum, which eventually give rise to an obliterative endarteritis, adventitial scarring, and patchy medial necrosis with destruction of elastic fibers. These destructive changes of the aortic wall produce diffuse or focal weakening, placing the patient at risk for aortic root dilatation and aneurysm formation. Contraction of scar tissue in the proximal aorta may lead to coronary ostial stenosis.

The predilection for involvement of the proximal aorta has been attributed to the large number of vasa vasorum in the ascending and transverse arch⁴. The aforementioned pathologic changes are responsible for the three major forms of symptomatic cardiovascular syphilis: aortic regurgitation, coronary ostial stenosis, and saccular aneurysm. The time from onset of the primary infection to formation of an aneurysm may vary from 15 to 30 years⁵. These aneurysms can occur anywhere in the aorta, but most commonly are found in the ascending aorta or transverse aortic arch. Most syphilitic aneurysms are saccular.

Once a syphilitic aneurysm is formed, its course is one of progressive enlargement until it either produces symptoms from compression on surrounding structures or death from rupture⁶⁻⁸. Aneurysms of the transverse arch may impinge on the trachea, esophagus or recurrent laryngeal nerve, resulting in stridor, dyspnea, dysphagia, hoarseness and pain.

The presence of a thoracic aneurysm can be

suspected from the plain chest radiography and confirmed with computed tomography. Diagnosis of tertiary syphilis can be confirmed with serology and pathology. The VDRL test may not be sensitive, but confirmation can be done with treponemal test such as treponemal pallidum particle agglutination (TPHA). Pathologic examination of the aortic wall will reveal an inflammatory infiltrate within the adventitia consisting of lymphocytes and plasma cells distributed predominantly around the vasa vasorum.

Once the diagnosis is confirmed, surgery should be recommended. The operation can be performed under total circulatory arrest with antegrade cerebral perfusion. Results after surgery are generally good.

CONCLUSION

Syphilitic aortic aneurysms are rarely seen today, but they are occasionally seen in developing countries. Because it takes a very long time for these diseases to progress until they manifest clinically, their occurrences should be kept in mind.

REFERENCES

1. Brockbank EM. Syphilis and aneurysm. *BMJ* 1925;2:606-7.
2. Jackman JD Jr, Radolf JD. Cardiovascular syphilis. *Am J Med* 1989;87:425-33.
3. Kent ME, Romanelli F. Reexamining syphilis: an update on epidemiology, clinical manifestations, and management. *Ann Pharmacother* 2008;42:226-36.
4. Kampmeier RH, Morgan HJ. The specific treatment of syphilitic aortitis. *Circulation* 1952;5:771-8.
5. Duncan JM, Cooley DA. Surgical considerations in aortitis. Part 3: Syphilitic and other forms of aortitis. *Tex Heart Inst J* 1983;10:337-41.
6. Heggveit H. Syphilitic aortitis. A clinico-pathologic autopsy study of 100 cases, 1950-1960. *Circulation* 1964;24:346-55.
7. Leung JSM, Mok CK, Leong JCY, et al. Syphilitic aortic aneurysm with spinal erosion. *J Bone Joint Surg Br* 1977;59-B:89-92.
8. Fulton JO, Zilla P, De Groot KM, et al. Syphilitic aortic aneurysm eroding through the sternum. *Eur J Cardiothorac Surg* 1996;10:922-4.