ID: JCDR/2012/4559:2451

Case Report

Internal Medicine Section

A Syphilitic Aortic Aneurysm: an old Friend Revisited

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ABSTRACT

An aortic aneurysm which is secondary to Syphilis, is a rare entity these days. A 45-year-old female presented with an insidious onset of breathlessness for evaluation. The patient was found to have a mediastinal mass. Contrast enhanced computed tomography (CECT) showed the presence of an aortic aneurysm with an intra-mural clot. Compression atelectasis of the left lung and a moderate left pleural effusion were also noticed. The VDRL and the TPHA tests were done, since this morphology of an aneurysm on the radiographic imaging led to a suspicion of Syphilis .The patient was proved to be have Syphilis, which was the aetiological cause of aneurysm in this case. The patient succumbed due to a massive haemoptysis, within one week of the diagnosis. The present report highlights the need to test every suspected syphilitic aortic aneurysm with both the VDRL and the TPHA tests, since the former alone are likely to give false negative results in late Syphilis. Also, this report highlights the importance of an early intervention in such cases, which may otherwise prove to be fatal.

Key Words: Saccular aneurysm, Syphilitic aneurysm, Aortic regurgitation, Volume rendered image

INTRODUCTION

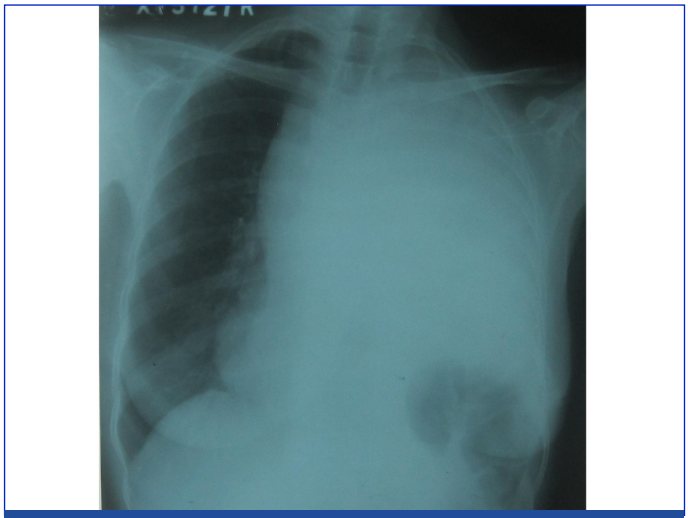
An aneurysm is defined as an abnormal focal dilatation of the blood vessels. An aortic aneurysm in a middle aged individual commonly results from inflammatory or infective causes. Cardiovascular diseases which include aortitis and aortic aneurysms occur in about 10% of the patients with untreated Syphilis, usually 10-30 years after the primary infection. Syphilitic aneurysms are seen in 2% of the HIV negative patients [2]. Without a surgical treatment, the mortality rate at 1 year, can reach up to 80%, due to high rates of rupture of these aneurysms [12] .We are reporting the occurrence of syphilitic aortic aneurysm with a large peripheral thrombus in a middle aged female.

CASE REPORT

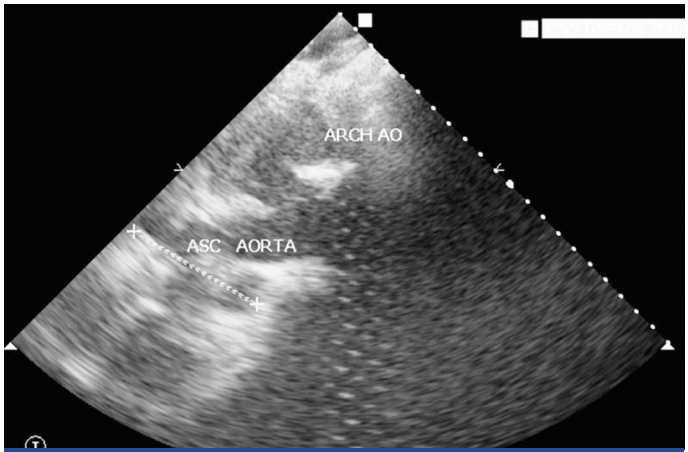
A 45-year-old female presented with the complaints of insidious onset breathlessness, a dull aching chest pain on the left side and dry cough for the preceding three months. She developed progressive breathlessness since the past 10 days. She was not known to have Diabetes mellitus or hypertension and there was no history which was suggestive of trauma, tuberculosis or connective tissue disorders in the past. Her husband was a lorry driver and he had expired 10 years back, the cause of which was not known to the patient. She had 2 children.

The general physical examination revealed a poorly built woman with a BMI of 20 and lymphadenopathy (3 firm nontender nodes of 2*1cm) in the posterior triangle of the neck. Her chest examination revealed diminished movements of the left hemi thorax. The left hemithorax was dull on percussion. The breath sounds were absent on the left hemithorax. The cardio vascular examination revealed a blowing early diastolic murmur in the aortic area.

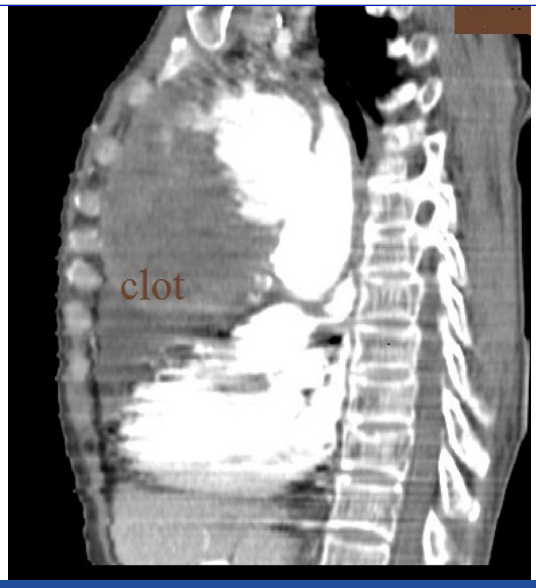
The routine investigation of the blood revealed a high erythrocyte sedimentation rate (ESR) of 80mm in the first hour. The sputum smear examination for acid-fast bacilli (AFB) was negative. The Mantoux test was non-reactive. FNAC of the lymph nodes showed a reactive hyperplasia without giant cells. Her electrocardiogram was normal. She tested negative for HIV/AIDS, Hepatitis B and



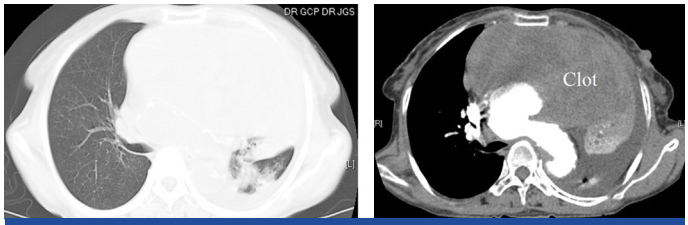
[Table/Fig-1]: Posteroanterior chest X-ray demonstrates unilateral homogenous opacification of left hemi thorax with mediastinal shift elevated left diaphragm as shown by high placement of gastric bubble.



[Table/Fig-2]: 2D echocardiography in the parasternal long axis view showing dilated ascending aorta and arch.



[Table/Fig-3]: Contrast enhanced CT scan showing mediastinal mass contiguous with aorta and enhancing intensely (small arrow). Left lung shows compression atelectasis (large arrow).



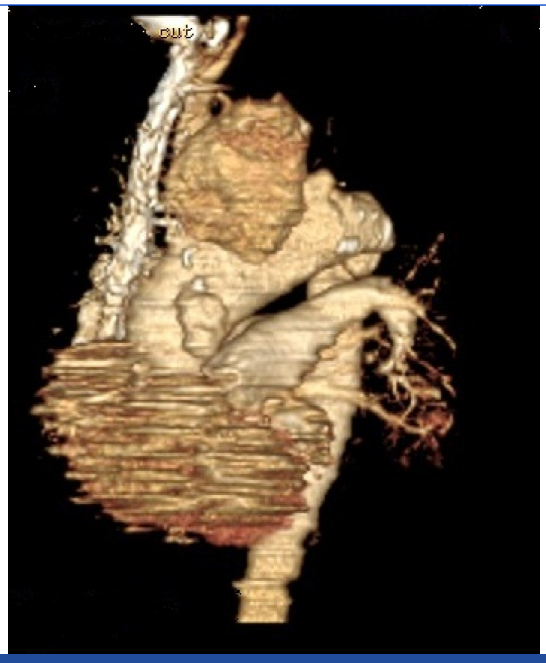
[Table/Fig-4]: CT angiography shows a large peripheral intramural thrombus measuring 13.9*9cm (arrows). Sagittal cut (4a), transverse cut (4b).

Hepatitis C. The Venereal Disease Research Laboratory (VDRL) test was negative.

Her chest radiograph showed a homogenous opacification of the left hemithorax, with an apparent shift of the mediastinum and perihilar calcified lymph nodes [Table/Fig-1].

Ultrasonography of her chest showed a 10cm × 8cm sized mixed echogenic lesion with internal hyperechoic floating echoes and a moderate, left sided pleural effusion. Diagnostic thoracocentesis revealed a bloody fluid which was negative for malignant cells and she had Adenosine deaminase levels of 20IU. Colour Doppler echocardiography revealed a dilated ascending aorta (aortic root of 6.4cm), moderate aortic regurgitation , mild pericardial effusion and good left ventricular and right ventricular functions [Table/Fig-2].

Non-contrast computer tomography of the chest showed a large mediastinal mass which was contiguous with the aorta which showed soft tissue attenuation. Contrast enhanced computer tomography of the chest showed a large out pouching over the anterior superior aspect of the ascending arch of the aorta. A majority of the outpouching was occupied by a soft tissue dense lesion, with a postero-central residual lumen communicating with the aortic lumen. The soft tissue material within the aorta showed significant enhancement and it produced a mass effect with a partial collapse of the left lung, a displaced aorta, the superior vena cava and the pulmonary artery. The outpouching measured 13.7*10.1 centimetres. There was moderate pleural effusion with an elevated left dome of the diaphragm, which were suggestive of compression of the phrenic nerve [Table/Fig-3]. For a better depiction of the aneurysm, a CT angiography was done. The CT angiography



[Table/Fig-5]: Volume rendered image reconstruction of the aneurysm showing minimal atherosclerotic features.

showed a giant saccular aneurysm with a maximum diameter of 13.9 × 9cm, which arose from the ascending aorta and the arch of the aorta, with an eccentric mural thrombus and a postero central residual lumen, which communicated with the aortic lumen. The abdominal aorta was normal in calibre [Table/Fig-4]. Most parts of the aneurysm contained a thrombus. Mild cardiomegaly and a pericardial effusion could be noted. The volume rendered (VR) images of the aneurysm were produced by using computer software [Table/Fig-5].

The computer tomographic images showed no significant atherosclerotic or dystrophic lesions in the aneurysm, thus suggesting a probable non atherosclerotic aetiology. The images showed the normal thickness of the aortic wall and this ruled out inflammatory arteritis as a probable aetiology .The bicuspid valves were of normal morphology. The VDRL test was repeated again and this time, it showed a low titre of 20:1. The Treponema Pallidum Haemagglutination Antibody (TPHA) titres were found to be positive, with a titre of 1:800 (the titres of 1:80 and above are significant). ELISA, which was done to test for HIV was negative.

A diagnosis of late Syphilis which was complicated by a syphilitic aortic aneurysm was made and the patient was given a course of benzathine penicillin, 2.4 million units per week for three weeks and tab metoprolol 12.5mg 1-0-0. She was advised to approach a cardio-thoracic surgeon for a surgical management at the earliest. She expired 10 days later due to a massive haemoptysis.

DISCUSSION

These days, syphilitic aortic aneurysms are a rare entity, since effective treatments are available for this infectious disease. Only few case reports have been published on this disease in the literature over the last decade. In a study which was done on untreated Syphilis, 10% of the patients were found to develop cardiovascular Syphilis after an incubation period of 10-30 years [1,2,5], 16% had gumma formation and 7% had neurosyphilis. The cardiovascular complications of Syphilis include syphilitic aortitis, aortic aneurysms, aneurysms of the sinus of the valsalva and aortic regurgitation. An aneurysm is defined as the pathological dilatation of the segment of a blood vessel. An aneurysm of the aorta is said

to be present when the diameter at any site is 5cm or more. The signs and symptoms vary according to the size and the location of dilatation. Frequently, the patients are asymptomatic. The other features include hoarseness of the voice, backache, haemoptysis, dysphagia, superior vena cava obstruction, a pulsatile mass and sternal erosions.

A giant aneurysm of the thoracic aorta in a young HIV seronegative individual in the absence of a history, which is related to atherosclerosis, trauma or Marfan's syndrome, is unusual and when such a lesion is detected, a further diagnostic work up is required to confirm an infection which is caused by Syphilis. In a recent case-control study, the prevalence of cardiovascular Syphilis in HIV seropositive individuals was found to be 14.3% and the prevalence of the same in HIV sero-negative individuals was two percent [2].

Syphilitic aortitis causes focal destruction of the media, with loss of the elastic smooth muscle fibres and scarring. This leads to an aortic dilatation and an aneurysm. The most common sites of these TAAs are the ascending thoracic aorta (36% of the cases), followed by the aortic arch (34% cases), the proximal descending thoracic aorta (25% cases), and the distal descending thoracic aorta (5% cases). Aortic sinus involvement occurs in less than 1% of the cases and it is most often asymmetric [9]. In late Syphilis, the non-treponemal tests like the VDRL test and the rapid plasma reagin test are less sensitive (70%-75%) as compared to the treponema specific tests such as TPHA, the micro haemagglutination assay and the fluorescent treponemal antibody absorption test (94%-95%) [3].

The imaging of an aortic aneurysm is best done with CECT of the chest and CT angiography. The MRI and MR angiography are slightly superior to CT in the evaluation of an aortic dissection. Morphologically, the syphilitic aneurysms are saccular but about 25% are fusiform. A pencil-thin dystrophic aortic wall calcification is found in up to 40% of the patients [10]. This is most severe in or is entirely confined to the ascending aorta. The delicate calcification is frequently obscured by the thick, irregular, coarse calcification of secondary atherosclerosis. In the cases of arteritis, the wall of the aneurysm is frequently thickened.

The location of an aneurysm can provide clue to its cause. In a study on 249 aneurysms of the aorta and its branches, which was done by Fomon et al., [12], it was found that the involvement of the ascending aorta alone was usually associated with annuloaortic ectasia, In contrast, atherosclerosis is a more diffuse process and it rarely involves only the ascending aorta.

The definitive treatment of an aortic aneurysm is surgical repair, which involves resection of the dilated portion of the aorta and its replacement with a synthetic vascular graft [14]. The simultaneous presence of an aortic regurgitation or a significant coronary disease must also be treated at the same time [14]. A more recent and modern procedure is percutaneous endovascular stent grafting, which has a lower mortality rate [4].

Without a surgical treatment, the mortality rate at 1 year can reach up to 80% due to the high rate of the rupture of these aneurysms [11,12].

CONCLUSION

The present report highlights the need to test every suspected patient with an aortic aneurysm for Syphilis with both VDRL and TPHA since the former alone are likely to give false negative results in late Syphilis. It also emphasizes the need of an early surgical intervention since the mortality is very high in such cases.

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FINANCIAL OR OTHER COMPETING INTERESTS:

None.

Date of Submission: **May 18, 2012**
Date of Peer Review: **Jul 03, 2012**
Date of Acceptance: **Aug 16, 2012**
Date of Publishing: **Sep 30, 2012**