Syphilitic heart - a case report

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Abstract

Heart and its great vessels are involved in tertiary syphilis in the form of syphilitic aortitis often 20 years or more after primary infection is contracted. In a 60 year old male cadaver, we found an enlarged heart with aneurysm of ascending aorta and semilunar septa was found protruding in to the lumen of arch of aorta distal to the origin of left subclavian artery. Right pulmonary vessels were dilated. Right and left atrial cavity were dilated, left ventricular hypertrophy with dilatation was noted. His pathological findings of the specimens were suggestive of syphilitic aortitis. Cardiovascular syphilis though uncommon is still a significant cause of mortality and morbidity, which can be reduced by adequate screening, accurate diagnosis and appropriate treatment.

Key words: aortitis, ascending aorta, arch of aorta, aneurysm, periarteritis, cardiomegaly.

Introduction

The heart is a hollow fibromuscular organ roughly pyramidal in shape placed within the pericardial sac in the middle mediastinum. It consists of four chambers, right and left atria (RA & LA) and right and left ventricles (RV & LV). RA receives venous blood from superior and inferior venae cava and coronary sinus. The blood enters the RV through right atrio-ventricular orifice, and thence ejected to pulmonary trunk (PT). The PT conveys deoxygenated blood from RV to the lungs. LA receives oxygenated blood through four pulmonary veins. From LA blood enters the LV through left atrio-ventricular orifice and ejected in to ascending aorta (AA) for systemic circulation. The AA begins in the aortic annulus, ascending upwards and to the right and continuing as arch of aorta (AOA), descending thoracic aorta and finally enters the abdomen as abdominal aorta through the aortic opening of diaphragm at T12 vertebral level.

The heart and its great vessels can be affected in various kinds of infectious disorders among which one of the most dangerous and life threatening disease would be involvement of heart in syphilis. The cardiovascular involvement appears in the tertiary stage of syphilis, often 20 years or more after primary infection.

This is a report of a case affected with syphilitic heart disease with cardiomegaly and aneurysm of its great vessels. The aim of this study is to make morphometric and morphological analysis of all the structures involved in this case and also studying the pathological changes and to emphasize the fact that cardiovascular manifestations are still a significant though uncommon cause of mortality and morbidity.

Case report

The present case was observed while dissecting a 60 year old male cadaver during dissection teaching for medical undergraduates in the anatomy department at M S Ramaiah Medical College, Bangalore. In this case we encountered an enlarged heart with abnormalities of great vessels. Various dimensions of the heart and great vessels were recorded, and the heart was opened along the flow of blood. The interior of the heart was observed in detail and the abnormalities were recorded and photographed and necessary measurements of the structures involved were noted. The tissue involved was sent for pathological examination.

Observation

Gross examination of the specimen revealed that heart was grossly enlarged suggesting cardiomegaly. AA
and AOA distal to the origin of left subclavian artery (LSA) were dilated. Lumen of this dilated part of AOA showed a semilunar septae projecting into the lumen. The right pulmonary vessels were found to be dilated but the left pulmonary vessels and coronary arteries were normal.

There was an additional branch arising from AOA between the origin of brachiocephalic trunk (BT) and left common carotid artery (LCCA), which was 3mm in diameter coursed forwards to supply the anterior part of the fibrous pericardium. [Fig. 1]

Fig. 1 shows anterior view of heart. 1. Dilated AA; 2. Extra branch from AOA; 3. BT; 4. LCCA; 5. LSA; 6. Dilated AOA, 7. Interior of 6 showing a semilunar septae.

**Abbreviations**: AA- Ascending aorta; AOA- arch of aorta; LCCA- Left common carotid artery; LSA- left subclavian artery.

The following measurements were recorded:
- Total weight of the specimen (heart with the vessels) - 750 grams
- Maximum transverse diameter of the heart - 14.8 cm
- Diameter of the heart from apex to base - 12.5 cm
- Maximum transverse diameter of AA - 6.6 cm
- Width of AOA proximal to the origin of BT - 7.2 cm
- Width of AOA distal to the origin of LSA - 4.8 cm
- Diameter of right pulmonary artery - 3.9 cm, left pulmonary artery - 2 cm.

Diameter of right pulmonary veins, upper - 3.2 cm, lower - 3.7 cm.
Diameter of left pulmonary veins, upper - 1.6 cm, lower - 1.3 cm.

After opening the heart along the flow of blood following observations were made: RA and LA cavity appeared to be dilated. Mitral valves were thickened and chalky white in appearance and the mitral orifice showed stenosis.

RV cavity and wall thickness appeared normal. LV cavity was dilated and the wall thickness was increased (1.5 cm) suggesting eccentric hypertrophy. Aortic and pulmonary valves and coronary arteries were normal on visualization.

**Histopathological findings**: Myocardium shows no significant pathology, pericardium showed periarteritis and endarteritis with lympho plasma cell infiltration. Sections from aorta showed atherosclerosis with superimposed thrombus with evidence of organization. Right and left pulmonary arteries showed evidence of periarteritis and fibrosis. The right and left pulmonary veins showed periarteritis, and endarteritis. Right and left coronary arteries and branches of AOA were normal. [Fig. 2]

Fig. 2 Photomicrograph showing perivascular lympho plasmocytic infiltration (red arrow) within the pericardial fat (black arrow).
Discussion

All the findings like cardiomegaly, aneurysm of ascending aorta, histopathology findings and dilatation of right pulmonary vessels were suggestive of syphilitic heart disease with isolated right pulmonary vessel aneurysm.

Syphilis is caused by Treponema pallidum subspecies pallidum is the microaerophilic spirochete that causes syphilis, a chronic venereal disease with multiple clinical presentations. The disease progresses through primary, secondary, and tertiary phases. Involvement of the cardiovascular system is the most dangerous sequel of the tertiary phase. This usually manifests as syphilitic aortitis (SA) which leads to slowly progressive dilation of the aortic root and arch which causes aortic valve insufficiency and aneurysm of ascending aorta².

In the natural course of cardiovascular syphilis, the primary infection is followed by T. pallidum invasion of the aortic wall within the adventitia and the lymphatic vessels. The rich lymphatic system in the AA is one of the main reasons for the tropism of spirochetes. The vasa vasorum later undergoes a process of endarteritis obliterans, necrosis of medial layer and infiltration of plasma cells³.

There have been relatively few surveys of syphilitic heart disease reported in the literature over the past 10 years. Studies of large series of necropsies are worthwhile to detect trends in incidence, changing patterns of lesions, and the influence of therapeutic measures. In clinicopathological autopsy of 100 cases, they divided the Syphilitic heart disease into five categories: uncomplicated SA, syphilitic aortic aneurysm, and syphilitic aortic valvulitis with insufficiency, and syphilitic coronary ostial stenosis and syphilis of the myocardium. In this series 36 cases were considered to be uncomplicated, the remaining 64 cases exhibited complications like aortic aneurysm, aortic insufficiency and coronary ostial stenosis⁴.

In a case report, a 57 year old patient presented with neurosyphilis and a large ascending aortic aneurysm was detected on CT angiogram. The pathogenesis of SA has been recognized for decades and its pathologic features are well documented. SA is reported in 70 - 80 % of untreated cases after the primary infection and in 10% of these patients significant cardiovascular complications will occur such as aortic aneurysm, aortic regurgitation and coronary ostia stenosis. The AA is the segment most commonly affected (50 %), AOA (35%) and the descending aorta (15%)⁵.

The post mortem incidence of syphilitic heart disease out of 4173 autopsies performed at the Ottawa General Hospital from 1950 to 1964 was reported to be only twenty six cases that satisfied the general accepted morphological criteria of SA. Decreasing incidence, changing patterns of lesions, frequent failure to diagnose is to be considered⁶.

Madke et al also reported a case of middle aged man, who presented with a pulsatile swelling on the left side of the anterior chest wall with gradual increase in size. He had past history of unprotected sexual intercourse and gave history of ulceration on glans penis 20 years back. CT angiography showed aneurysm of AOA and AA. In view of his past history, positive serology and imaging studies, he was diagnosed to be having luetic aortopathy. This case was reported to be an extreme rarity in the present antibiotic era⁷.

In European and North American countries, degenerative and dystrophic diseases represent the bulk of ascending aorta pathology, few cases of post traumatic aneurysm or non infectious aortitis, are also reported. Syphilitic aneurysm has practically vanished in recent large series, but is still present in many countries with inadequate health policies and alterations in severity and course of syphilis in immunodeficient patients may occur⁸.

The variations of branches arising from aortic arches are well known and documented by several authors in
different case reports. Adachi classified branching pattern of arch of aorta as three types. Type A is the classical branching pattern in to BCT, LCCA and LSA which is seen in 80% of the individuals. Type B (11%) is the common trunk incorporating the LCCA and BT leaving only two branches originating from the AOA. Type C has the left vertebral artery as fourth branch. Number of aortic visceral branches like bronchial, esophageal, pericardial, mediastinal arteries has been explained by Bergman and others as observed in the present case.

Conclusion

It has been estimated that cardiovascular syphilis is responsible for 10 - 15 % of all heart disease presenting after age 50. Syphilis still remains a real threat, and until it is eradicated, it will continue to cause clinical and subclinical cardiovascular disease. The purpose of this report is to emphasize the fact, that the cardiovascular manifestations of syphilis are significant cause of morbidity and mortality. Any patient with aortic aneurysm, aortic insufficiency and coronary ostial stenosis should be subjected to VDRL serology test to rule out syphilis. Hence death and complications due to syphilis can be prevented with adequate screening, accurate diagnosis and definitive treatment at an early stage.

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References


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