

Case Report

Heller-Döhle Syndrome: A Rare Case of Syphilitic Aortitis with Calcified Ascending Aortic Aneurysm in an Elderly Lady

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Abstract

Background: Tertiary syphilis with cardiovascular complications are a rarity in the antibiotic era in developed nations although diagnoses of syphilitic aortitis is not uncommon in developing countries. It usually presents in the 4th to 5th decade of life and is unusual in the elderly. Aortic aneurysms and aortic regurgitation due to aortic root dilatation is the usual presentation. Nevertheless, it can remain undetected for the duration of its latent period which may extend upto 40 years and is often diagnosed on routine evaluation.

Presentation of Case: We report a case of an 82 year old south indian lady who presented with clinical features of aortic regurgitation who was found to have a large calcified ascending aortic aneurysm with no neurological manifestations.

Conclusion: We discuss the diagnostic challenge due to the unusually long latent period in our patient, the epidemiologic concerns, treatment options and possible complications.

Keywords: Syphilis; Syphilitic aortitis; aortic aneurysm; Heller Dohle syndrome

Peer Reviewers: Imtiaz Ahmed Wani, MD, District Hospital, Directorate Health services, Kashmir, India; Shagufta Tahir Mufti, MD, Faculty of Medicine, King Abdulaziz University, Saudi Arabia

Received: June 10, 2013; **Accepted:** July 23, 2013; **Published:** August 6, 2013

Competing Interests: The authors have declared that no competing interests exist.

Consent: We the authors confirm that our patient and his family have given informed assent/consent for the publication of this case report.

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Introduction

Syphilis is one of the most well-known sexually transmitted diseases known to man for

the past several centuries with such varied manifestations that it was dubbed “the great imitator” by the father of modern medicine, Sir. William Osler [1]. It is caused by the spirochete *Treponema Pallidum*. It can present

in one of four different stages: primary, secondary, latent and tertiary. It may also present congenitally. Transmission is primarily by sexual contact and by vertical transmission. The late manifestations of syphilis (Tertiary Syphilis) involve the nervous system, the heart or the skin and subcutaneous tissues (gummas). However, the incidence of these late manifestations of syphilis have been reduced to a rarity since the era of antibiotics. Before the discovery of penicillin, tertiary syphilis infection was the most common cause of thoracic aortic aneurysm, resulting in up to 10% of cardiovascular deaths [2]. Syphilitic aortitis 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 ostial stenosis [3]. The ascending aorta is the segment most commonly affected (50%) and the rich lymphatic arrangement in the ascending aorta that may predispose greater mesoaortitis is believed to be the cause for larger involvement of this segment [4]. Cardiovascular syphilis usually manifests in the fifth decade of life, typically 10–40 years after the primary infection. It is uncommon to come across such cases which present beyond the 7th decade of life. Without surgical treatment, the mortality rate at 1 year can reach 80% due to the high rate of rupture of aneurysms [5]. The authors report an unusual case of syphilitic ascending aorta aneurysm with an aortic regurgitation in an elderly lady of 82 years of age.

Case Report

An 82 year old lady with no premorbidities presented to us on an out patient basis with history of progressive exertional dyspnoea and palpitations. Initial clinical evaluation revealed a high volume collapsing pulse and an early diastolic decrescendo murmur in the aortic area suggesting a diagnosis of aortic incompetence. Her nervous system examination was

essentially normal. Initial blood investigations were unremarkable except for a mild anemia with a haemoglobin of 10.2g/dL. The ELISA test for HIV-1 came back negative. The chest X-Ray (Fig.1) revealed a large aneurysm of the ascending aorta extending to the aortic root with evident calcification of the aneurismal wall. A trans-thoracic echocardiogram showed moderate to severe aortic regurgitation with aortic root dilation and moderate left ventricular hypertrophy. There were no other significant valve lesions nor any intra-cavitary masses. A CT of the brain showed only age related atrophy.

In this clinical scenario, in spite of the advanced age of the patient and lack of a history of high risk behaviour, the diagnosis pointed to tertiary syphilis with cardiovascular involvement in the form of syphilitic aortitis and ascending aortic aneurysm. There was no evidence of neurological involvement. The patient then tested positive for Venereal Disease Research Laboratory (VDRL) and Treponema Pallidum Particle Agglutination (TPHA) tests done on her blood. She had been sexually inactive for over 30 years and denied any high risk sexual behaviour. Added to the fact that the patient denied any high risk behaviour she also had no recollection of any primary lesions or symptoms of secondary syphilis in her past. She was started on intravenous crystalline penicillin which was continued for 14 days and started on long term cardiac medications on which her symptoms improved. The option of surgical repair of the aortic aneurysm was discussed with the patient and her relatives which they decided not to undergo. She was also not willing to undergo a coronary angiogram to look for ostial stenosis. She was discharged and lost for follow up subsequently. The unusual presentation of this patient is mainly due to her late presentation as well as a lack of neurological involvement in the face of a probable latent period of 30 years or more. Another feature of note was the large aneurysm in the ascending aorta which seemed to be of a stable nature with no evidence of

imminent rupture. This was attributed to the significant calcification seen on the walls of the aneurysm.

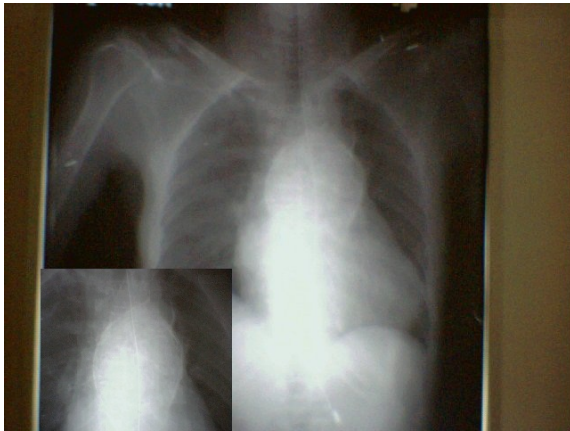


Figure 1

Discussion

Syphilis may manifest at any of its four stages, namely, Primary, Secondary, Latent or Tertiary. In the antibiotic era, prevalence of syphilis has come down drastically, especially in the developed countries. In India, the public health problem posed by syphilis is still significant. Transmission of the disease is primarily by heterosexual contact. A subset of cases are due to vertical transmission during pregnancy. The organism, *Treponema Pallidum* is highly infective and an individual inoculated with an average of just 57 organisms has a 50% chance of being infected [6]. Since the spirochete can pass through both compromised skin and intact mucous membrane, it may be transmitted by kissing near a lesion as well as oral, vaginal and anal sex [6]. Around 30-60% of individuals exposed to primary or secondary syphilis will contract the disease. The prevalence of syphilis in India ascertained by various studies range from 10-15% of all sexually transmitted diseases [7-9]. However, studies published in the state of Kerala where our patient resides show a slightly higher rate of syphilis among the STDs as compared to the rest of the country [9, 10]. Tertiary syphilis consists of three possible manifestations which are

Cardiovascular, Neurological and involvement of subcutaneous tissue (Gummas). Tertiary syphilis may involve any one of the above or may present as a combination of two or more system involvements. Our patient had features of isolated cardiovascular tertiary syphilis.

Tertiary syphilis with cardiovascular manifestations has nearly disappeared in developed countries with the advent of modern anti microbial therapy, although it is still not uncommon in developing nations. Syphilitic aortitis, eponymously called Heller-Döhle syndrome, is the most common manifestation of syphilitic cardiovascular involvement. The primary lesion of cardiovascular syphilis is aortitis, an inflammatory response to the invasion of the aortic wall by the *Treponema pallidum* that evolves to obliterative endarteritis of the vasa vasorum and results in necrosis of the elastic fibres and connective tissue in the aortic media. The resulting weakening of the aortic wall will progress into the late vascular manifestations of syphilis [3, 4]. Recent studies show an increase in cases reported related to HIV infection [11-13]. The diagnosis is often missed due to the rarity of this entity in the 21st century. In late syphilis, non-treponemal tests like VDRL test and rapid plasma reagin test have decreased sensitivity (71-73%), when compared with treponema-specific tests such as TPHA, Microhaemagglutination test, fluorescent treponemal antibody absorption test (FTA-ABS) which have sensitivity ranging from 94-96% [14]. According to Kuramochi et al. [15], syphilis proved with serology is necessary to make the diagnosis of syphilitic aortitis as the histologic findings of mesoaortitis by itself is not diagnostic. In the presence of an aortic aneurysm, syphilitic serological testing is advised. Screening is also advised for certain high risk individuals and groups [16]. CT angiogram is considered the best imaging study to define the anatomy and size of the aneurysm, but in the setting of an aneurysm, the echocardiogram and coronary angiogram are mandatory to exclude aortic

regurgitation and coronary flow-limiting lesions and coronary ostial stenosis [2, 4]. Surgical repair is the definitive treatment of aortic aneurysm, which involves the resection of the dilated portion of the aorta and replacing it with a synthetic vascular graft [17]. If there is an associated aortic regurgitation or significant coronary disease they should preferably be surgically treated at the same time. Even so, surgery and specific antibiotic treatment does not exclude future manifestations of the disease, even after eradication of the organism, which makes permanent follow-up needed [18]. The chances of relapse are especially high in HIV co-infected patients [19]. Also, there is a re-emergence of syphilis among high-risk individuals particularly among men having sex with men (MSM) and commercial sex workers. This changing epidemiology may mean that we may see the delayed neurological and cardiovascular complications of tertiary syphilis in increasing frequency in the times to come [20]. In our patient, in view of history of sexual inactivity for over 30 years, there are several postulates as to how she may have contracted the infection. The most likely possibility is that the initial transmission may have taken place close to 40 years prior to presentation and may have been transmitted by her husband, but the initial symptoms may have been disregarded. Such long latent periods, though rare, is not unheard of. This underlines the need for a high index of suspicion in such cases for proper identification and management.

Conclusions

Though tertiary syphilis is a rare condition among developed nations, the incidence is still quite high in the developing countries where primary syphilis goes untreated and indeed even undetected quite often. The diagnosis of cardiovascular syphilis can often prove elusive and frequently diagnosed only after irreversible damage has taken place. Tertiary syphilis with

cardiovascular manifestations have significant morbidity and mortality risk with the possibility of aneurismal rupture, aortic dissection and acute myocardial infarction due to ostial stenosis a likely prospect. Hence, the diagnosis of tertiary syphilis should be kept in mind in all cases of ascending aorta aneurysms and aortic regurgitation due to root dilatation. A high index of suspicion must be maintained if the patient belongs to a group at high risk of developing sexually transmitted diseases.

Acknowledgements

The authors would like to acknowledge the patient who consented to publication: written informed consent was taken from the patient for publication of this case report and the accompanying images.

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