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Tertiary Syphilis Presenting with Abdominal Pain and Aortitis

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Abstract

Context*: There are four stages in which syphilis progresses. It is important to note that an aneurysm involving the upper abdominal or the descending thoracic aorta occurs in about 10-15% of cases of cardiovascular syphilis.*

Case Report*: We present a case report about a 59-year-old male who presented with abdominal discomfort and was later diagnosed with tertiary syphilis and aortitis with aneurysmal dilation of the infrarenal aorta. He was surgically treated with an endovascular repair of an aortic aneurysm. His cerebral spinal fluid resulted positive for VDRL, and the patient was treated with IV Penicillin.*

Conclusion*: Proper diagnosis and antibiotic treatment are critical when syphilis is suspected.*

Introduction

Given the significant extent and impact of syphilis in the US, the National Syphilis control campaign was launched in 1938 by US Surgeon General Thomas Parran. The campaign focused on serologic testing, public education, and the treatment of syphilis. By the mid-1900s, there was nearly a 75% decrease in the reported cases of syphilis, and by 1975, there was a decrease by nearly 90% of syphilis cases reported. However, in recent years, the World Health Organization (WHO) has estimated that the yearly incidence of syphilis has increased to nearly 12 million. (1)

At the turn of the twenty-first century, there has been a steady increase in the number of syphilis cases. This likely resulted from demographic and behavioral changes and was notably higher amongst men who have sex with men (MSM) in the early 1980s. In the late 1980s, there was an increased association between those who used crack cocaine and heterosexuals. By the early 2000s, again, there had been a reported increase rate of syphilis among MSM. Notably, they also had a higher rate of coinfection with HIV. (1)

In addition to sexual transmission, syphilis may be acquired vertically from an infected pregnant woman to her unborn fetus, in addition to infected needles and very rarely by tattooing. Syphilis has been referred to as “the great imitator” due to its vast clinical presentations. Characteristically, there are four stages in which syphilis progresses, including the following: primary syphilis, secondary syphilis, latent syphilis, and lastly tertiary syphilis.2 It is essential that if suspected, even if the individual is asymptomatic, the diagnostic confirmation and proper treatment are started.

Case Report

A 59-year-old male with a past medical history significant for hypertension (HTN), transient ischemic attack (TIA) in 2008, patent foramen ovale (PFO), left atrial appendage thrombus diagnosed on transesophageal echocardiography (TEE) in 2008, type 2 diabetes mellitus, tobacco use, and Hepatitis B present with a two-day history of abdominal discomfort. The pain was stabbing in quality and rates the pain a 9 out of 10. It emanated from his lower midline abdominal pain and was a constant stabbing feeling. It was exacerbated with exertion and had no ameliorating factors. Over-the-counter analgesia provided no relief.

While in the emergency department (ED), he was given IV hydromorphone, which provided complete relief. Of note, the patient stated that he had similar symptoms two years ago. He was diagnosed with rectal Gonorrhea secondary to rectal intercourse, for which he received ceftriaxone and doxycycline. Over the past two decades, the patient related he had dozens of sexual partners and 2 in the past year.

His family history revealed no history of autoimmune disease, aortic aneurysms, or arteritis. He did have a pertinent family history of cardiovascular disease. His father had several cardiac stents placed and ultimately succumbed to his cardiovascular disease at 60. He is a corporate lawyer and currently has a very high-stress job where he feels he is on the brink of being fired. Upon exam, mucous membranes were moist without lesions, has anicteric sclera, and Argyle Robertson's pupil was challenging to assess. His cardiovascular exam revealed a regular rate and rhythm, warm extremities, and distal pulses 2+. Bloodwork revealed CRP 45.1, ESR 36, WBC of 7.6, and screening syphilis serology was positive. Confirmatory testing was ordered, and the rapid plasma reagin (RPR) titer was 1:1024. He was started on weekly IM benzathine penicillin.

An abdominal CT was ordered and revealed the multifocal atherosclerotic plaque of the aorta and major abdominal arteries, as seen in Figure 1. Aneurysmal dilation of the infrarenal aorta measured up to 33 mm secondary to a focal outpouching along the left aspect of the mid aorta measuring up to 12 x 17 x 15 mm axial imaging. It extended beyond the aortic wall compatible with penetrating atherosclerotic ulcer. There was surrounding stranding and multiple prominent retroperitoneal lymph nodes. The two right renal arteries had severe stenosis of the anterior artery appreciated, and severe stenosis of the proximal left renal artery was noted. There was normal patency caliber on the superior mesenteric, splenic, portal, and hepatic veins.

He underwent a successful endovascular repair of the aortic aneurysm with three separate stents under fluoroscopy. Follow-up imaging showed patency of the renal and iliac vasculature. While awaiting confirmation, the patient started related to increased forgetfulness and difficulty concentrating. This raised the concern for Neurosyphilis, and a lumbar puncture was performed on hospital day 7. Increased WBC and protein were noted, the CSF return positive for VDRL, and based on CDC recommendations, the patient was started on IV Penicillin hospital day 7 of admission along with two weeks of prednisone. The patient received IV penicillin for a total of 28 days. Of note, the patient's HIV test came back negative. He was to follow up to treat his Hepatitis B. Hepatitis B virus DNA was positive (891 IU/mL, 8.95 log IU/mL), Hepatitis E antigen was also positive, and Hepatitis C was negative.

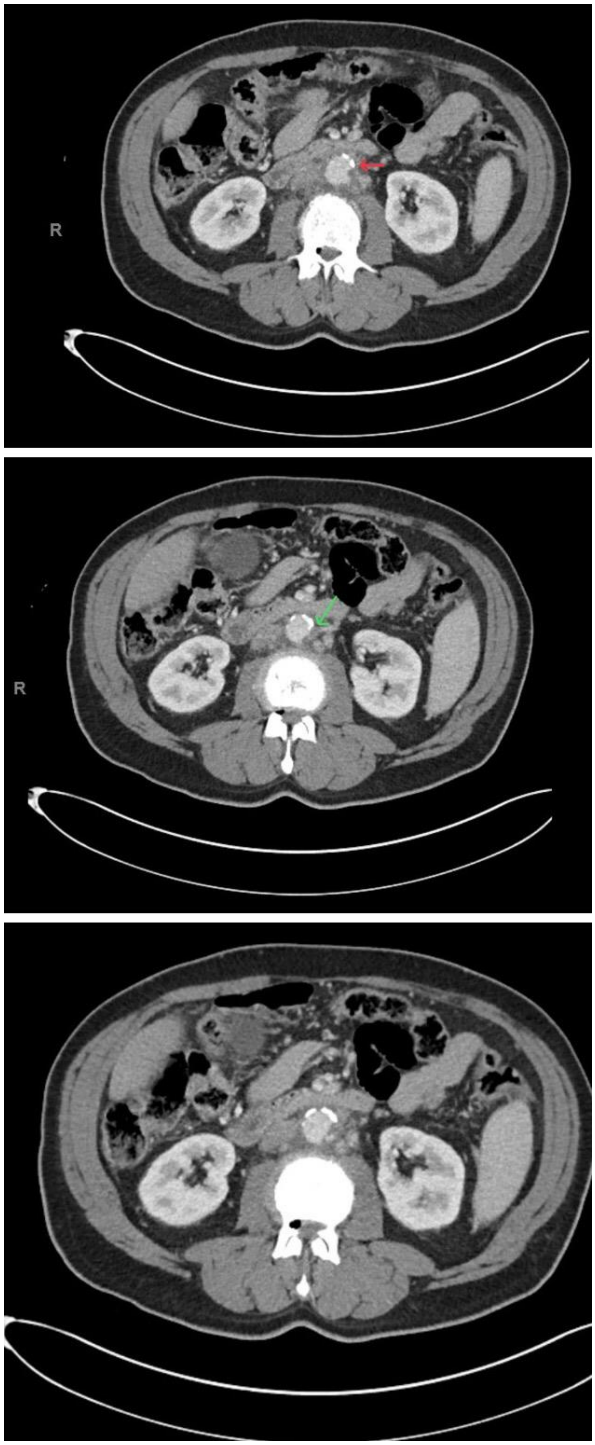

Figure 1: Axial abdominal CT revealed multifocal atherosclerotic plaque (Red arrow) of the aorta and major abdominal arteries, as seen in aneurysmal dilation of the infrarenal aorta measured up to 33 mm secondary to a focal outpouching along the left aspect of the mid aorta measuring up to 12 x 17 x 15 mm axial imaging (Green arrow). It extended beyond the aortic wall compatible with penetrating atherosclerotic ulcer.

Discussion

Syphilis, an STI, is a disease caused by a spirochete; Treponema Pallidum was described as early as the 15th century. Characteristically, there are four stages in which syphilis progresses, including the following: primary syphilis, secondary syphilis, latent syphilis, which is divided into the early stage and late stage, and finally tertiary syphilis. Primary syphilis has an incubation period anywhere from 10-90 days. Lymphadenopathy is notable near the inoculation site and typically manifests with a painless indurated ulcer, also known as a chancre, that generally is one to two cm in diameter. Without treatment, it will take nearly a month and a half for the macrophages to destroy most spirochetes present, but not all of them. Secondary syphilis typically results from the systemic spread of the spirochetes and may be present for weeks to months after initial inoculation. In this stage, a generalized mucocutaneous rash and lymphadenopathy occur. Other manifestations of secondary syphilis include condylomata lata, anterior uveitis, optic neuritis, nephritis, and mucosa lesions in the mouth and genital tract. (2)

Early latent syphilis typically refers to the period between the end of secondary syphilis and is present less than one year from inoculation. Late latent syphilis refers to infection being present greater than one year. Most individuals are asymptomatic and will have a positive serological test. However, it has been noted that nearly 25% of patients may develop recurrent secondary manifestations if left untreated. It is estimated that almost 2/3 of patients with latent syphilis will remain in this latent stage. However, 1/3 of those that remain untreated in the latent stage will develop tertiary syphilis. Some of the most notable characteristics in this stage include cardiovascular and neurological manifestations and scattered granulomatous lesions, also known as gummas. (2)

Treatment with antibiotics has significantly decreased the number of cases that progress to tertiary syphilis. Cheng et al. note there has been a decline because of the efficacy in antibiotic treatment and an increase in public health awareness of early stages of syphilis. Even with these efforts, there are still documented cardiovascular syphilis cases occurring. Cardiovascular manifestations of tertiary syphilis typically present anywhere from 10-20 years after the initial infections. (3) Of note, concerning the presentation of cardiovascular syphilis, aortic aneurysm is a relatively rare type. (4)

Cardiovascular syphilis is divided into three main categories: the great vessels, medium-sized arteries, and the heart. Involvement of the aorta can present with any of the following: aortic aneurysm, uncomplicated aortitis, ostial coronary stenosis, and aortic regurgitation. The involvement of the aorta varies depending on the location. Nearly 50% involve the ascending aorta, 35% involve the arch, and only 15% involve the descending aorta.5 It is important to note that an aneurysm involving the upper abdominal or the descending thoracic aorta is relatively rare. In 90% of these cases, the RPR will test positive, as seen in our patient. (5)

Duncan and Cooley explain that cardiovascular syphilis can be divided into five categories as follow: uncomplicated syphilitic aortitis, syphilitic aortic aneurysm, syphilitic aortic valvulitis with aortic insufficiency, syphilitic coronary ostial stenosis, and finally syphilitic myocarditis. Uncomplicated syphilitic aortitis typically occurs in 70-80% of the untreated cases, and if left untreated or unrecognized, 10% of these patients will further develop complications. A syphilitic aortic aneurysm is mainly saccular, and less commonly, fusiform. They may have various clinical presentations depending on the surrounding structures near the aneurysm. Moreover, although not diagnostic, linear calcifications are common on CT. Operative treatment in addition to antibiotics is crucial and holds relatively low mortality. Syphilitic aortic valvulitis with aortic insufficiency typically involves the enlargement of the aortic root and may dilate the aortic valve annulus. If this occurs, regurgitation will be present. On imaging, these leaflets will not contain calcifications if it is a solitary syphilis infection. Syphilitic coronary ostial stenosis can develop from the encroachment of the ascending aorta onto the coronary ostia. Treatment is then targeted on bypassing any of the obstructed coronary arteries in addition to aortic valve replacement. (6)

Furthermore, when there is clinical suspicion for aortitis, it is vital to obtain a detailed family and social history. This is important because although aortitis typically results from Takayasu's disease, mycotic aneurysms, and syphilis, other diseases with known and unknown etiologies can also present with the same clinical manifestation. For example, rheumatic fever, Giant-cell arteritis, and systemic lupus erythematosus are some of the few that may all develop aortitis. Additionally, Del Re et al. notes that it's substantially uncommon for patients to present with aortic aneurysms due to tertiary infection in the modern era because of the liberal use of antibiotics. (7) However, there should be an emphasis on the diagnosis and appropriate antibiotic therapy in suspected cases. (8)

Given our patient's history, in addition to the laboratory values and imaging, syphilitic aortitis was presumed and treated. Unfortunately, extensive use of transcatheter technologies has had its limitations when aortitis is suspected. This is mainly due to the lack of pathology confirmation via surgical specimens. In this case, histopathologic confirmation could not be obtained because of the endovascular approach and thus provides some limitations.

In conclusion, cardiovascular syphilis complicated by syphilitic aortitis and infrarenal abdominal aneurysm, although rare, should be diagnosed and treated promptly.7 Although patients may remain asymptomatic and without clinical manifestation for many years, the risk of further involvement is imminent. Treating the syphilis infection with sufficient antibiotics before surgery and post-operatively can decrease the development of additional circulatory and neurological symptoms. Additionally, a lumbar puncture is essential to rule out tertiary syphilis, which requires IV Penicillin.

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