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INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/14889
DOI URL: http://dx.doi.org/10.21474/IJAR01/14889

RESEARCH ARTICLE

CARDIOVASCULAR SYPHYLIS PRESENTING AS AORTIC ANEURYSM, AORTIC CALCIFICATION AND AORTIC REGURGITATON - A RARE ENTITY IN THE ANTIBIOTIC ERA - A CASE REPORT

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Manuscript Info

Manuscript History
Received: 10 April 2022
Final Accepted: 14 May 2022
Published: June 2022

Key words:-
Tertiary Syphilis, Aorta, Aneurysm, Regurgitation, Calcification

Abstract

Aortic aneurysm formation is a known complication of late syphilis. Here we report a case of37 year old female presented with breathlessness and chest pain.On evaluation ascending aorta aneurysm, descending aorta calcification and aortic regurgitation are identified, later on diagnosed to have syphilis. The diagnosis of the aortic aneurysm due to tertiary syphilis may be challenging due to deceptive clinical presentation and rarity of the disease because of widespread use of antibiotics for the management of earlier stages of syphilis.

Introduction:-

The first outbreak of syphilis recorded in 1495 in Europe during a French invasion and therefore called as French disease [1]. The term syphilis came in to use in 1530 by Italian physician named *Giralamo Fracastoro* and causative organism T.pallidum identified in 1905[2]. First treatment was started in 1910 with salvasran followed by penicillin use in 1943[3].

The incidence of late manifestations of syphilis have declined almost to a rare entity since the era of antibiotics. Following the introduction of penicillin therapy in the 1940s, the number of cases of syphilis of all stages declined to 95% in 2000[4].However, the number of cases of primary and secondary syphilis increased again in 2001, and the rate has continued to climb almost every year since that time. By 2018, the overall number of reported primary and secondary syphilis cases was 35,063, with a rate of 10.8 cases per 100,000 population.From 2013 to 2018 the rate of primary and secondary syphilis in women increased by over 170 percent. In 2018, the rate among women was 3 cases per 100,000 females [5].

Cardiovascular syphilis generally manifests in the fourth to fifth decade of life, around 15-30 years after the initial infection and the majority of patients remain asymptomatic [6]. Involvement of the ascending arch of the aorta and aortic valve is a consequence of vasculitis of the vasa vasorum ("endarteritis obliterans") caused by syphilis.

Case Report

A case of 37-year-old married non diabetic and non-hypertensive female came to ER with the chief complaint of shortness of breath chest pain for 6 months and palpitations for 6 months with significant past history and no family history. On examination there is high volume pulse with wide pulse pressure, apex beat shifted downward and outward and on auscultation there is loud A2 with early diastolic murmur. Complete hemogram and other

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hematological investigations were unremarkable, viral markers tested for HIV, Hepatitis B and C were negative. On x-ray radiopaque lesion seen along the descending aorta from the origin of arch of aorta, on HRCT aortic wall calcification involving root, arch of aorta, and descending aorta up to the level of D10 vertebra. Echocardiography showed ascending aorta ectasia with aortic root diameter measuring about 5.0cm with severe aortic regurgitation.The patient tested positive for treponemal pallidum particle agglutination (TPHA) on blood and positive for Rapid plasma regain (RPR) test on blood (titer 1/32). Cerebrospinal fluid examination is unremarkable.

In light of the aneurysm size and progressive nature of the patient's symptoms, surgical repair was recommended and penicillin therapy was started but unfortunately patient had lost to follow up and could not be traced further.

Figure 1:- 3D Reconstruction image showing aortic clacification.

Figure 2:- CXR PAV showing wall calcification involving aorta.

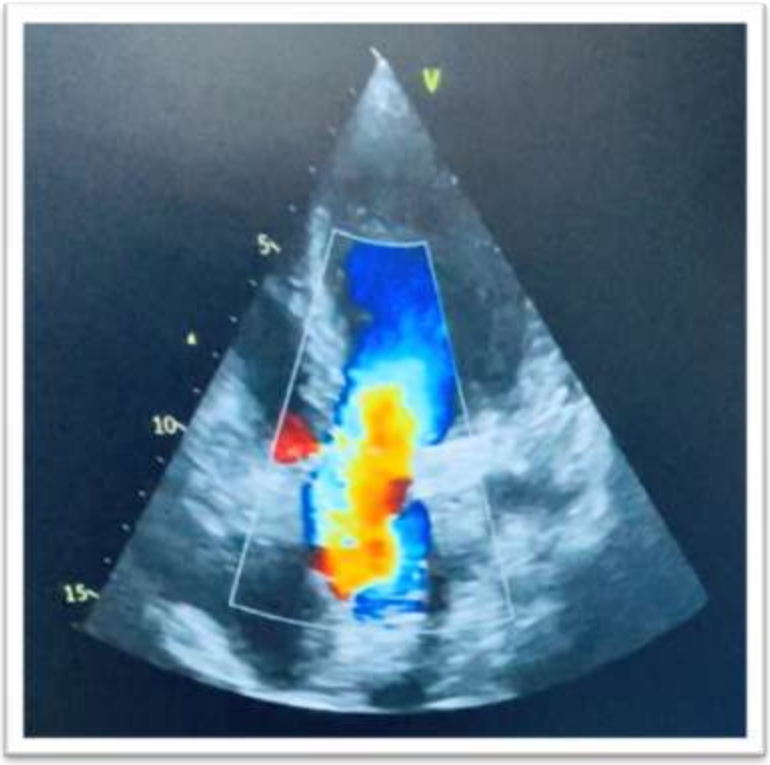

Figure 3:- 2D Echo showing severe aortic regurgitation.

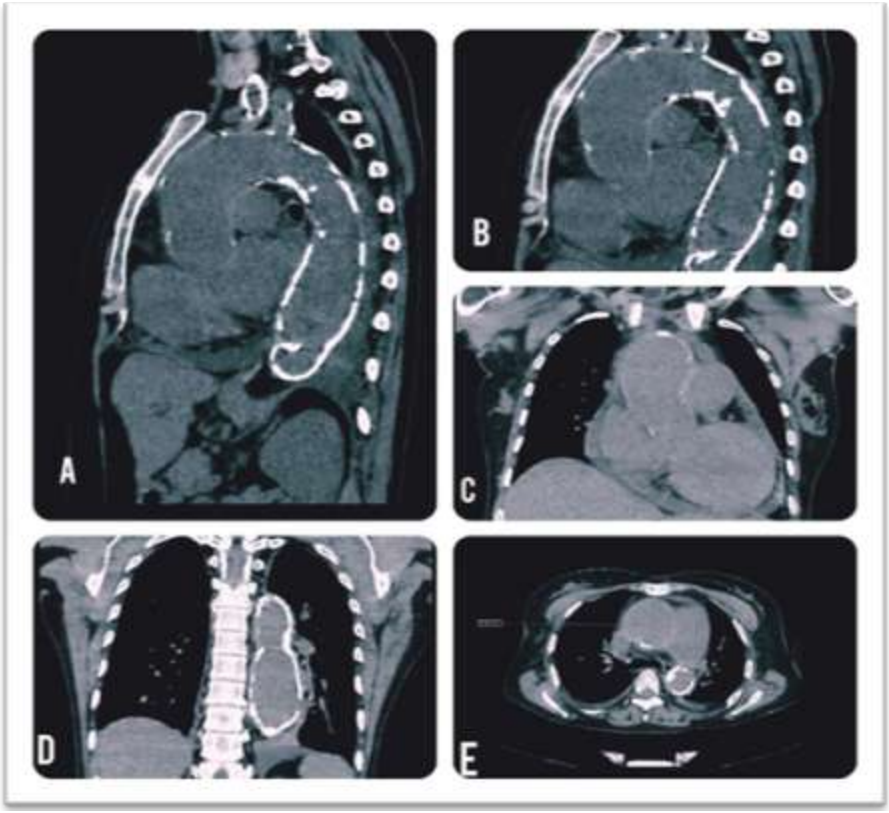

Figure 4A&B):- Sagittal section of CT thorax(mediastinal mode) , 4C&D) Coronal section of CT thorax (mediastinal mode) , 4E) axial section of CT thorax(mediastinal mode) showing ascending aortic aneurysm.

Discussion:-

In the preantibiotic era, asymptomatic aortitis was considered to be highly prevalent; in fact, it is said that the majority of patients with long-standing syphilis have subclinical aortitis undetectable on radiographs or at clinical examination [7]. The incidence of late manifestations of syphilis have declined almost to a rare entity since the era of antibiotics.However, the number of cases of primary and secondary syphilis increased again in 2001, and the rate has continued to climb almost every year since that time. As an example, 8724 cases were reported in 2005. From 2011 to 2015, there was a 67 percent increase nationwide in primary and secondary syphilis rates [8]. By 2018, the overall number of reported primary and secondary syphilis cases was 35,063, with a rate of 10.8 cases per 100,000 population [9].Here we report a case of young female with fusiformascending aortic aneurysm, descending aortic calcification and severe aortic regurgitation. Cardiovascular syphilis classically involves thoracic ascending aorta (50%) because predominant lymphatic enrichment of this segment predisposes to mesoaortitis. The aortic arch (35%) and descending segment of the aorta (15%) are less commonly involved [10]. Uncomplicated syphilitic aortitis is the most common (70-80%) manifestation of cardiovascular syphilis and 10% of these patients develop fusiform or saccular aortic aneurysm, aortic regurgitation, and stenosis of the coronary ostium [11]. . The rarity of this etiology makes the diagnosis difficult, mainly because syphilis testing is not routinely used. In the presence of an aortic aneurysm, particularly in younger patients, syphilitic serological testing is advised [12]. In late syphilis, non-treponemal tests like VDRL test and rapid plasma regain (RPR) test are less sensitive (71–73%), when compared with treponema-specific tests such as TPHA, micro-hemagglutination test, fluorescent treponemal antibody absorption test (94–96%) [13].The definitive treatment of aortic aneurysm is surgical repair, which involves resection of the dilated portion of the aorta and replacing it with a synthetic vascular graft. The simultaneous presence of aortic regurgitation or significant coronary disease should be surgically treated at the same time [14].The postoperative treatment with benzathine penicillin, in doses recommended for tertiary syphilis, is regularly implemented to decrease the chances of relapse. However, it is known that even with the elimination of T. pallidum, the chance of recurrence of the disease still persists. Because of this, there is a need for a long-term follow-up of the patient [15]. After diagnosis of syphilis was made, she was started on benzathine penicillin G 2.4 million units intramuscular weekly for 3 weeks and advised for surgical repair of aortic aneurysm but unfortunately, we lost follow up of the patient.

Conclusion:-

Atypical presentation of this typical disease as cardiovascular syphilis in not uncommon even in this antibiotic era,hence high-risk individuals with a thoracic aortic aneurysm should be tested for syphilis screening followed by the treponemal specific test as early diagnosis and treatment of tertiary syphilis with antibiotic therapy and subsequent surgical repair of syphilitic aortic aneurysms can prevent fatal complications and mortality.

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