Syphilitic aortitis: About a case in South Colombia

Aortitis sífilítica: a propósito de un caso del sur colombiano

Diego J. Alvis-Peña^{1*}, Carlos H. Calderón-Franco², Julieth Gonzales-Cerón³, María P. Alvis-Peña⁴, and Yexica A. Rodríguez-Rodríguez⁵

¹Department of Internal Medicine and Research, Hospital Universitario Hernando Moncaleano Perdomo, Neiva, Huila; ²Department of Internal Medicine and Research, Clínica Mediláser, Florencia, Caquetá; ³Department of Internal Medicine and Research, Hospital Universitario Hernando Moncaleano Perdomo, Neiva, Huila; ⁴Academic Unit, Universidad Surcolombiana, Neiva, Huila; ⁵Academic Unit, Fundación Universitaria Navarra, Neiva, Huila. Colombia

Abstract

The case of a 73-year-old female patient obtained and resident of Florencia, Caquetá (Colombia), with a history of hypertension who is admitted to a highly complex hospital due to a 3-month clinical picture of constant evolution in pain is presented. Thoracic oppressive type pain in the anterior region of the left thorax, non-irradiated, of intensity 8/10. The echocardiogram shows calcified aortic valve disease with severe aortic stenosis, aortic regurgitation, left ventricular hypertrophy, and mild grade mitral regurgitation. Within the pre-surgical studies, she presented VDRL with 1:4 dilutions and the diagnosis was confirmed with the result of the treponemal test and other infectious diseases were ruled out. Valvular replacement was performed with a bioprosthesis plus a supra-coronary valvular tube. In the biopsy, chronic scar valvulitis with extensive calcifications was confirmed. After the surgical intervention, discharge was performed due to adequate clinical evolution.

Key words: Aortitis. Syphilis serodiagnosis. Syphilis cardiovascular.

Resumen

Se expone el caso de una paciente de 73 años procedente y residente de Florencia, Caquetá (Colombia), con antecedentes de hipertensión arterial, que ingresa a un hospital de alta complejidad por un cuadro clínico de 3 meses de evolución consistente en dolor torácico tipo opresivo en la región anterior izquierda del tórax, no irradiado, de intensidad 8/10. En el ecocardiograma se evidencia enfermedad calcificada de la válvula aórtica con estenosis aórtica grave, insuficiencia aórtica, hipertrofia del ventrículo izquierdo e insuficiencia mitral leve. Dentro de los estudios prequirúrgicos presentó VDRL con 1:4 diluciones, se confirmó el diagnóstico con el resultado de la prueba treponémica y se descartaron otras enfermedades infecciosas. Se realizó remplazo valvular con bioprótesis y tubo valvular supracoronario. En la biopsia se confirmó la valvulitis crónica cicatricial con extensas calcificaciones. Posterior a la intervención quirúrgica se dio egreso por adecuada evolución clínica.

Palabras clave: Aortitis. Diagnóstico serológico de la sífilis. Cardiovascular syphilis.

Correspondence:

*Diego J Alvis-Peña

E-mail: diego-927@hotmail.com

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Introduction

Given that syphilis is a disease caused by *Treponema pallidum*, which is transmitted by the sexual, hematogenous or trans-placental routes, characterized by successive stages of evolution, and that its treatment with penicillin is so far well-known and efficacious, it should not be a public health problem, but current figures show the opposite since, in Colombia, 6754 people were diagnosed with syphilis in 2010, out of whom 836 had a late syphilis diagnosis and, in 2011, there were 992 cases with late syphilis diagnosis¹. The high proportion at outpatient services brings to light the existence of barriers to its adequate classification according to its stage, as well as the lack of opportunity and quality of the care provided to patients who seek medical attention for a sexually-transmitted infection².

Cardiovascular syphilis is the hallmark of aortitis, which is a consequence of tertiary syphilis, caused by the spirochete infection that affects aortic segments with inflammation and subsequent scarring, producing necrosis of the middle layer, as well as fibrous proliferation of the tunica intima³. Clinical manifestations can occur as aortitis, aortic aneurysm, aortic regurgitation, or coronary ostial stenosis, with aortic regurgitation being the most common complication⁴. Latency periods are long, ranging from 10 to 30 years after primary infection in patients who did not receive treatment or with incomplete treatments⁵. Although the prevalence of cardiovascular syphilis after the penicillin era has relatively declined, cases continue to be reported in developing countries⁶.

Several cases of syphilitic aortitis-type cardiovascular complications have been described, which were surgically managed due to aortic aneurysmal dilatation associated with severe aortic regurgitation, with favorable results being obtained^{4,5}.

Case presentation

Seventy-three-year-old woman, of mestizo ethnicity, native to and resident of Florencia, Caquetá (Colombia), housewife. She was admitted to the emergency department of a high-complexity hospital of Neiva (Colombia) due to a 3-month history of oppressive chest pain at the left anterior region of the chest, non-radiating, 8/10 in intensity, which increased with medium-effort daily activities, and that was associated with progressive dyspnea within the previous 15 days and improved with rest. Important previous history data included hypertension, hysterectomy 15 years prior, and cholecystectomy 20 years prior. She denied relevant

family history conditions. She was on treatment with metoprolol, amlodipine, acetylsalicylic acid, losartan, atorvastatin, bisacodyl, and alprazolam.

On initial physical examination, she had blood pressure of 140/80 mmHg, heart rate of 72 beats/min, respiratory rate of 18 breaths/min, pulse oximetry of 92% inspiring ambient air; she was afebrile, weight was 59 kg, hydrated without jugular venous engorgement, normally expandable chest, rhythmic heart sounds with Grade III systolic murmur audible in all foci with predominance of the aortic focus, with no presence of edema, and neurological system without alterations.

Echocardiogram showed severe aortic stenosis with an opening of 0.84 cm and peak velocity of 4.52 m/s, with moderate aortic regurgitation lateralized toward the anterior mitral leaflet, left ventricular walls concentric hypertrophy due to chronic systolic overload and ascending aorta dilatation.

Coronary arteriography showed coronary arteries without lesions and aortic valve with moderate calcification. Chest computed tomography angiography confirmed the ascending aorta aneurysmal dilatation, of 48 mm in diameter (Fig. 1A and B).

Laboratory tests at admission showed no alterations (hemoglobin 12.6 g/dL, hematocrit 38.4%; platelets 188,100/mm³; prothrombin time 17.3 s; and partial thromboplastin time 39.9 s); electrolytes were within normal ranges (sodium 145.6 mmol/L; potassium 4.15 mmol/L), and troponins were negative. Since there were no laboratory alterations, and in view of the described findings, cardiovascular surgery was scheduled for aortic valve replacement plus supracoronary tube versus Bentall. Infectious disease testing reported bacilloscopy without acid-fast bacilli being found in 100 observed fields, hepatitis B surface antigen 27,18 (positive), hepatitis C surface antigen 27,86 (positive), anti-HCV IgG 2,32 (positive), negative human immunodeficiency virus, and reactive VDRL (Venereal Disease Research Laboratory) in 1: 4 dilutions. In view of these findings, surgery was put on hold and hepatitis C viral load, hepatitis B anticore IgM, hepatitis B antigen, hepatitis B antibodies, hepatitis B viral load, and treponemal test were determined; with the infectious disease tests results, decisions could be made regarding the surgical procedure of the patient. The treponemal test report was positive, and treatment with benzathine penicillin was, therefore, started at a dose of 2,400,000 IU by the intravenous route, weekly, for 3 weeks.

Hepatitis B and C were ruled out, since viral load results reported no infection (<10 copies/mL for both and anticore IgM 0.050, not reactive).

Figure 1. Tomographic view **A:** and three-dimensional reconstruction **B:** there is evidence of significant ectasia of the aortic ascending segment, with a diameter of 48 mm, and transverse diameter of the descending segment is 23 mm, with presence of parietal atheromatous plaques without intimal dissection.

Figure 2. **A:** Surgical intervention by sternotomy with surgical correction of aortic valve replacement with a 21-mm Perimount bioprosthesis plus 30-mm supracoronary tube being performed. **B:** post-surgical chest X-ray.

Given that this was a case of cardiovascular syphilis, treatment with benzathine penicillin was administered and, after the last dose, surgery was carried out. Surgical strategy was discussed taking into account the computed tomography, transthoracic echocardiography, and coronary angiography reports. Aortic valve replacement with a 21-mm Perimount bioprosthesis plus 30-mm supracoronary tube was decided (Fig. 2A and B).

Surgically-resected material biopsy reported chronic scarring valvulitis with extensive calcification.

The patient post-operative evolution was uneventful. After the surgical procedure, consultation was requested with social work and clinical psychology to evaluate her family support network, which was adequate, and after 10 post-operative days with an excellent recovery, she was discharged on treatment with carvedilol

(12.5 mg/12 h), amiodarone (20 mg/24 h), losartan (50 mg/12 h), and acetylsalicylic acid (1 mg/24 h).

Discussion

We present the case of a 73-year-old woman with cardiovascular syphilis and aortitis with aortic valve compromise.

Syphilis is a chronic systemic bacterial infection that can last for decades, caused by the *T. pallidum* spirochete, subspecies *pallidum*. The only host is the human being. Disease presentation can vary and mimic different clinical manifestations, which is why it is known as “the great imitator.” Diagnosis is based on non-specific lipid serological detection (rapid plasma reagin [RPR] and VDRL), followed by specific treponemal serologic tests^{7,8}. A recent increase in the incidence of this disease has been observed all over the world, and its cause is multifactorial, which includes increased high-risk and unprotected sexual activity, migration, and social changes, which reduce opportunity and accessibility to health services for early and timely treatment. However, treatment regimens are currently well defined and successful⁹, with first line of treatment being penicillin, which was used in the patient before the surgical intervention at a dose of 2,400,000 IU by the intramuscular route, weekly, for 3 weeks.

Tertiary syphilis can occur in approximately one third of untreated patients and cause granulomatous lesions, called gummata; other manifestations include cardiovascular and neurological conditions. At present, it is rare for this type of manifestations to be found due to the success of antibiotics, but this infection can develop in the long-term (in years and perhaps decades) after an initial infection without proper management and can be observed in up to 70% of untreated patients^{4,5,9,10}.

Syphilitic aortitis is the cardinal manifestation of cardiovascular tertiary syphilis. It is secondary to vasa vasorum vasculitis, which generates obliterative endarteritis with ischemic lesion of the tunica media of aorta, as well as loss of smooth muscle and extracellular matrix. Compromise of the ascending thoracic aorta has been more commonly found, due to the significant amount of vasa vasorum and extensive lymphatic network of this region, which is invaded by *T. pallidum* during primary infection. The exceptional involvement of the thoracic aorta is due to the absence of abdominal aortic vasa vasorum. From another perspective, aortic wall weakening leads to aortic root dilatation and potential associated complications, with the most important being aortic valve regurgitation (as in our patient), syphilitic aortic aneurysm, and coronary ostial stenosis¹⁰⁻¹³. Thoracic aortic aneurysms grow upward at a rate of 0.10 cm/year¹⁴. Consensus

guidelines advise surgical evaluation regardless of size for all patients with symptoms suggestive of aneurysm expansion, as well as for asymptomatic individuals with an ascending aorta sinus diameter of 5.5 cm or more¹⁵. Aneurysms are complicated due to invasion and erosion of the chest wall, which further complicates the case at the time of surgical intervention.

As previously mentioned, the most common complication is aortic regurgitation, which occurs in 20-30% of patients and is secondary to aortic root dilatation. In addition, there are symptoms of heart failure, and it is usual for a murmur to be auscultated at the right sternal border¹⁶; in our patient, a Grade III systolic murmur was audible in all foci, with high predominance in the aortic focus. The second most common complication is syphilitic aneurysm, which is 3 times less frequent than aortic regurgitation and is detected in 5-10% of patients; Grade II aortic regurgitation, moderate grade aortic valve calcification, and aneurysm of the aortic root and ascending aorta were found in our patient, which is consistent with the literature on the most common location of aneurysms (50% in the ascending aorta, 35% in the aortic arch and 15% in the descending aorta)¹⁷⁻²⁰.

Cardiovascular syphilis diagnosis is based on serological, histological, and imaging criteria and, in our patient, it was established under these three premises. The first initial non-treponemal test was confirmed with treponemal-specific serological tests. In addition, it was confirmed with echocardiography, percutaneous angiography, and CT angiography with three-dimensional reconstruction. On histological examination, the ascending aorta arterial wall was observed with a degeneration of the tunica media characterized by an accumulation of small basaloid deposits without inflammation and with irregular and transmural areas of disorganization and separation of the fibers. In the aortic valve wall, thickening and sclerosis of the spongy layer was observed, with large areas of calcification. A mild sub-epithelial chronic inflammatory infiltrate was observed on the ventricular surface, with focal erosion and fibrin aggregates. No granulomas or microorganisms were found in the evaluated material. These findings are suggestive of what is reported in the literature as a consequence of fibrous thickening of the adventitia with vasa vasorum loss, surrounded by lymphoplasmacytic cells and loss of elastin fibers in the media, with intima media atherosclerotic thickening, and generally without evidence of spirochetes within the aorta^{7,8,10}. As for antibiotic treatment, it was with penicillin due to its adequate sensitivity and because it is the first therapeutic line; dosing was carried out according to the regimen established by the guidelines.

The presented case offers one of the best solutions for patients with this pathology and who are of high risk and may be the best evidence that it is an available treatment option for the management of this infection and its life-threatening complications.

Conclusion

Syphilis is an infection that continues to be prevalent in our population and, for this reason, it is highly important for health personnel to become familiar with its broad and different clinical manifestations, which include cardiovascular presentations, with aortic valve insufficiency, ascending aortic aneurysm, and coronary ostial obstruction being more common, which manifest themselves according to the natural evolution of the disease at tertiary syphilis stage.

Sexually-transmitted diseases, including syphilis, continue to be a public health problem in our region and in developing countries. Although cardiovascular syphilis is a rare and long-standing complication, human-talent health personnel should be able to recognize its early stages, its broad clinical manifestations, and its risk factors in order for early diagnosis to be established, taking into account both non-treponemal and treponemal tests, as well as timely and comprehensive treatment, and thus being able to reduce possible complications.

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Conflicts of interest

The authors declare that they have no conflicts of interest.

Ethical disclosures

To carry out the research, ethical aspects of the Declaration of Helsinki and resolution 8430 of the Ministry of Health of Colombia were taken into account^{21,22}, with this research being classified at a minimum-risk level.

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this research.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

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