Syphilitic aortitis as a rare cause of coronary ostial stenosis

Aortitis sifilitica como causa rara de estenosis de los ostium coronarios

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To the Editor,

This is the case of a 62-year-old man who presented to the emergency department with signs of an acute neurological syndrome. He remained under regular monitoring due to spastic paraparesis. The patient’s past medical history also included dyslipidemia, active smoking, former alcohol abuse, and psoriasis. His routine medication included daily aspirin 150 mg, and simvastatin 20 mg. Due to severe worsening of his neurological status, he was admitted for further evaluation. After careful clinical evaluation, diagnosis of cerebellar and pyramidal syndrome in the neurosyphilis setting was achieved. Penicillin was started. During hospitalization, cerebral magnetic resonance imaging revealed the presence of a massive hernia at C4-C5 causing significant spinal cord compression. Decompressive surgery was advised. During hospitalization, he complained of chest pain. The ECG showed signs of sinus rhythm with sustained diffuse ST-segment depression and ST-segment elevation in aVR and V1. The transthoracic echocardiography showed a severely impaired left ventricular ejection fraction with severe hypokinesia of the apex, anterior, posterior, and lateral walls. The aortic root was mildly enlarged, but no flaps were seen. Due to refractory chest pain and progressively worsening hypotension, the patient was given unfractionated heparin (5000 IU) and underwent an emergency coronary angiography that revealed the presence of critical left main coronary artery ostial stenosis [videos 1 and 2 of the supplementary data]. No further lesions were identified. Due to the complexity of the lesion, percutaneous angioplasty under left ventricular assist device was advised. It was necessary to make a multidisciplinary decision due to the patient’s condition.

Due to the patient’s unstable and worsening hemodynamic condition, a coronary angioplasty using a drug-eluting stent was decided and successfully performed (figure 1, and figure 2). Before the angioplasty was performed, the patient was given a loading dose of ticagrelor 180 mg. The procedure was backed by intracoronary ultrasound (IVUS), which showed good stent positioning and expansion at the end of the procedure [minimum in-stent area of 16 mm²] [videos 3 and 4 of the supplementary data]. No signs of coronary artery dissection were reported. After the procedure, the patient was pain-free, and blood pressure levels came back to normal.

The transthoracic echocardiography was repeated, and confirmed a mildly dilated aortic root (40 mm to 41 mm) with apparent posterior wall thickening. The left ventricle was not dilated. The left ventricular ejection fraction was 30%-35% with an apical akinetic area, and anterior, lateral, and posterior walls. The right ventricular function was normal. No significant valvular disease, pericardial effusion or intracardiac masses were reported.

A thoracic computerized tomography scan showed multiple atheromatous aortic calcifications and significant wall thickening, which correlated to aortitis phenomena of syphilitic etiology. The patient remained on dual antiplatelet therapy and completed his antibiotic cycle with penicillin. The patient had favorable cardiovascular progression with gradual improvement of the left ventricular function and was discharged to the neurosurgery unit after 7 days. At 1 month, ticagrelor was withdrawn, and the patient underwent neurosurgery. His neurological recovery was uneventful and after 6 months, left ventricular function was normal.

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SUPPLEMENTARY DATA

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Although cardiovascular signs have been previously described in the medical literature as well-known complications of syphilis, this case illustrates a particularly rare cardiac complication in the modern era.1-3 A possible sign of syphilitic aortitis is ostial coronary narrowing that can lead to an acute myocardial infarction, most cases being identified post-mortem.1 The underlying mechanism can be associated with atherosclerotic plaques, inflammatory phenomena, and/or calcium protrusion to the coronary arteries.1-3 High level of suspicion and the proper clinical setting were essential to achieve diagnosis and further treatment.1 Other differential diagnoses can be questioned like ankylosing spondylitis, temporal arteritis, and Takayasu’s arteritis since they can cause ascending aortitis.2,3

This case underlines the complexity of treating ostial lesions of left main coronary artery especially in situations where coronary obstruction seems to be conditioned by calcifications at ascending aorta, aortic root or aortitis level. Displacement of calcium in the aorta can lead to critical obstructions that, in the emergency setting, can complicate percutaneous revascularization or make it unfeasible. The absence of left ventricular assist devices and the close availability of cardiac surgery in our hospital made this scenario frightening and difficult to manage in the acute phase. Former studies have reported angioplasties in patients with left main coronary artery ostial stenosis, most requiring left ventricular assist devices to support the angioplasty of left main coronary artery.4,6 Fortunately for the patient, emergence angioplasty was possible with favorable cardiovascular progression. The patient’s written informed consent was obtained.

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AUTHORS’ CONTRIBUTIONS
R. Flores, F. Mané, C. Braga, and C. Oliveira treated the patient. R. Flores drafted the manuscript, and F. Mané, C. Braga, and C. Oliveira reviewed it.

CONFLICTS OF INTEREST
None reported.

SUPPLEMENTARY DATA
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REFERENCES
To the Editor,

This is the case of a 57-year-old man with a past medical history of thoracic radiation in his adolescence in the context of Hodgkin’s lymphoma. The patient also has unclassified interstitial lung disease and endothoracic goitre with tracheal displacement creating a difficult airway. Back in 2014, the patient was diagnosed with chronic coronary syndrome and complete percutaneous revascularization of the right coronary artery was achieved. The echocardiograms revealed the presence of severe mitral valve stenosis due to valve calcification without signs of rheumatic disease, preserved systolic function, and severe pulmonary hypertension.

The patient started showing progressive signs of heart failure, and aortic valve replacement was indicated. Given the evidence of porcelain aorta, transcatheter aortic valve implantation (TAVI) was decided using a 23 mm Edwards SAPIEN valve (Edwards Life-sciences, United States) under deep sedation via femoral access. No complications were reported at discharge.

Follow-up in the valve clinic showed progressive worsening of dyspnea (New York Heart Association [NYHA] class III). Additional tests confirmed the progression of mitral stenosis. The transthoracic [TTE] and transesophageal (TEE) echocardiography confirmed the presence of extensive mitral annular calcification (MAC) causing severe mitral stenosis (mean gradient, 15 mmHg) without regurgitation, mild right ventricular dilatation with preserved function, and estimated systolic pulmonary artery pressure > 60 mmHg. The valve was working properly.

The case was reassessed, and the possibility of transcatheter biological aortic valve implantation in the mitral position was considered given the surgical and anesthetic risks involved.

The feasibility study of the procedure assessed using the 3mensio Structural Heart system (Pie Medical Imaging, The Netherlands) revealed the presence of mitral annular calcification with a circumferential extension of 298°, no calcification in the medial commissure, anteroposterior and intercommissural diameters of 24.5 mm and 33 mm, respectively, and a inner area of 646 mm². These measurements were considered eligible for the 29 mm Edwards SAPIEN valve implantation.

In the simulation, estimating the neo-left ventricular outflow tract (LVOT) area [340 mm²] was particularly relevant, considered low risk for LVOT obstruction [figure 1]. The results and the echocardiographic images obtained proved the case eligible for valve implantation in mitral annular calcification, also known as valve-in-MAC via transfemoral access. The patient was informed of the high complexity and morbidity and mortality associated with the procedure, with long-term results still unknown to this date.

The procedure was performed back in March 2022 with the patient presenting to the cath lab with clinical data of heart failure. Under general anesthesia and intubation with a fiberoptic bronchoscope, a conventional TEE probe was advanced uneventfully. Under TEE and fluoroscopy guidance, a transseptal puncture was performed via venous femoral. Afterwards, the left atrium was accessed with an Agilis Nxt catheter [Abbott Laboratories, United States], and the mitral valve was crossed using a conventional J-tip guidewire. With back-up from a 7-Fr coronary guide catheter, 2 Safari high-support guidewires [Boston Scientific, United States] were placed into the left ventricle. The interatrial septum was dilated using a 12 mm angioplasty balloon, and a 29 mm Edwards SAPIEN 3 valve was advanced until it was eventually placed inside the mitral orifice, and then properly aligned.