A Case of Undiagnosed Syphilis in a Patient Undergoing Coronary Artery Bypass Graft and Aortic Valve Surgery

Marcos M. Soliman, MD,* Ihab Dorotta, MD,* Steven P. Larosa, MD; Bruce W. Lytle, MD;†
Colleen Gorman Koch, MD, MS,*§ Judith A. Drazba, PhD;* E. Rene Rodriguez, MD;||
and C. Allen Bashour, MD*§

Abstract: We report a case of syphilitic aortitis suspected by the intraoperative appearance of the aorta. Diagnosis was confirmed by postoperative serological testing. Given the recent increase in the number of cases of primary syphilis, it is important to remain aware of the clinical and pathological manifestations of tertiary syphilis.

S yphilis is an uncommon but not completely eradicated disease. Introduction of antibiotics and the effort to increase public awareness of sexually transmitted diseases has led to a marked decrease in the incidence of syphilis in the United States and Western Europe. The latest outbreak of primary syphilis in the United States, which occurred in 1990 (50,000 cases) during the human immunodeficiency virus epidemic, brings to light the importance of health care providers remaining aware of the manifestations of latent syphilis. Tertiary syphilis develops in approximately 30% of untreated cases and can result in syphilitic aortitis and/or neurosyphilis. We present a case of syphilitic aortitis diagnosed in a patient who presented for aortic valve replacement and coronary artery bypass graft surgery.

CASE REPORT

A 61-year-old man was referred to our institution because of sudden onset of dizziness and vertigo and episodes of confusion and nausea for several months before admission. The patient had a medical history of gastroesophageal reflux disease, substance abuse, and sexually transmitted diseases. He admitted of drinking alcohol and denied smoking.

Physical examination revealed a grade 3 diastolic heart murmur at the heart base. Computed tomography of the head showed chronic small vessel ischemic disease. Carotid duplex ultrasound scanning revealed bilateral 20% to 39% stenosis in the internal carotid arteries. A chest radiograph showed a mildly dilated heart and tortuous aorta. Transthoracic echocardiogram demonstrated an ejection fraction of 50%, a trileaflet aortic valve with a moderately severe centrally directed jet of aortic insufficiency without a clear etiology, and a mildly dilated aortic root. Left heart catheterization showed a 40% ostial narrowing of the left main coronary artery, a 60% narrowing in the middle third of the left anterior descending coronary artery, and no calcification of the aortic valve. The patient was scheduled to undergo aortic valve replacement and coronary artery bypass graft surgery.

Intraoperative transesophageal echocardiography demonstrated a diffusely thickened aortic wall without atheroma (Figs. 1, 2). Because of the abnormal appearance of the aortic wall, the right axillary artery was used for cardiopulmonary bypass inflow cannulation. Cardiopulmonary bypass was initiated, and the heart was arrested with warm cardioplegic solution. The aorta felt soft and was cross-clamped, but it seemed abnormal, and the possibility of syphilitic aortitis was noted. The aortic root was enlarged to accommodate a size-23 Carpentier-Edwards bovine valve, and coronary artery bypass grafting was performed.

The patient’s postoperative course in the cardiovascular intensive care unit was complicated by confusion and prolonged initial intubation time. Serological confirmation of the diagnosis of syphilis was made: rapid plasma reagin (RPR) test was reactive at a dilution of 1:32, and the fluorescent treponemal antibodies–immunoglobulin G was positive. A lumbar puncture was performed to exclude neurosyphilis and demonstrated increased white blood cells (31 white blood cells predominantly lymphocytes [98%], protein of 81 mg/dL, glucose of 75 mg/dL, and a positive Venereal Disease Research Laboratory at a dilution of 1:8 [31]. Cerebrospinal fluid fluorescent treponemal antibody was also reactive consistent with the diagnosis of neurosyphilis. Human immunodeficiency virus serology was nonreactive.

Pathological examination of the aorta revealed heavy adventitial inflammatory infiltration predominantly by lymphocytes and macrophages with marked fibrosis, as well as vasulocentric inflammatory infiltration of the media predominately by plasma cells consistent with the diagnosis of syphilitic aortitis (Fig. 3). Intravenous antibiotic therapy was initiated with penicillin G. The patient’s confusion improved over the next several postoperative days, and he was extubated and had an uneventful recovery. He was discharged to home on a 14-day course of intravenous penicillin G therapy. A follow-up RPR test had been ordered but not obtained from the patient.

DISCUSSION

Tertiary syphilis is a slowly progressive inflammatory disease, which becomes clinically perceptible years after the
initial infection. Primary infection often goes unnoticed in situations where the painless ulcer is not secondarily infected, does not develop, or is present in an atypical location. A serological diagnosis becomes more difficult to make over time as up to 25% of patients will become RPR or Venereal Disease Research Laboratory negative. One third of the patient subset with untreated syphilis goes on to develop tertiary syphilis, and 7% and 10% of these patients may develop neurological and cardiovascular complications respectively. Sixteen percent of these patients develop characteristic gummas, which are soft gummy tumors composed of tissue that resembles granulation tissue. Cutaneous gummas may occur as ulcers or granulomatous lesions on the skin, or visceral gummas may present as a mass lesion in the central nervous system.

Cardiovascular complications of tertiary syphilis include (in decreasing frequency) the following: syphilitic aortitis, ascending aortic aneurysm, aortic valve insufficiency, and coronary ostial stenosis. Unlike atherosclerotic aortic aneurysms, which tend to occur more commonly in the descending aorta, syphilitic aneurysms more frequently affect the ascending aorta. These complications are usually diagnosed at postmortem.

Syphilitic aortitis results from the invasion of the aortic wall by the *Treponema pallidum*. These spirochetes appear in the adventitia of the aorta and travel into the media, probably

---

**FIGURE 1.** Chronic aortitis from the treponemal infection is manifested by thickening of the aortic wall in this midesophageal aortic valve long-axis view.

**FIGURE 2.** A color flow Doppler jet consistent with severe aortic regurgitation is displayed from this midesophageal aortic valve long-axis view. Note the mosaic of color beneath the aortic valve in the left ventricular outflow tract in diastole.

**FIGURE 3.** Light microscopic examination of the aorta. A, The Movat pentachrome stain shows the entire thickness of the aorta. The intima is seen in the upper part of the image and shows mild atherosclerosis. The media is clearly demarcated by the abundant elastic lamellae (black lines) that show some fragmentation and focal paucity. The lower half of the aortic wall is a markedly thickened and fibrotic adventitia (yellow) with inflammatory infiltrates around the vasa vasorum (Movat pentachrome). B, This micrograph shows very clearly the basophilic (blue) mononuclear infiltrates in the adventitia and the less prominent mononuclear infiltrates around the vasa vasorum that reach the media and intima (hematoxylin-eosin). C, High magnification around a small vasa vasorum shows mononuclear cells and plasma cells surrounding the vessel, which, in turn, is embedded in an area of dense fibrotic scar (hematoxylin-eosin). D, This micrograph shows the area framed in the rectangle in B. There is a dense mononuclear inflammatory infiltrate in perivascular location. From the small adventitial arteries, it is possible to see smaller branches that become the vasa vasorum as they travel into the fibrotic adventitia and media (hematoxylin-eosin).
through lymphatics in the vessel wall. Pathological characteristics include inflammation of the media around the vasa vasorum with infiltration by lymphocytes and plasma cells. The inflammation and infiltration lead to endarteritis obliterans and subsequent patchy necrosis in the media. Finally, the inflammation progresses to fibrosis both in the media and in the adventitia, with resultant weakening of the aortic wall and aneurysm formation. As in this case, the aorta often seems thickened because of fibrosis, and the obliteration of the vasa vasorum leads to focal scarring of the media, which, in turn, creates scars with a distinct grooved appearance resembling the superficial longitudinal grooves as in tree bark. Our patient had a thickened aorta and exhibited typical pathological manifestations of syphilitic aortitis but did not report history of primary syphilis.

A tree bark appearance of the adventitial surface of the aorta is not pathognomonic for syphilis and can be seen in connective tissue diseases including systemic lupus erythematosus, Marfan syndrome, and Takayasu aortitis. Patients with syphilitic aortitis may not necessarily have a history of primary infection, and serological testing can be negative in patients previously treated with penicillin.

In contrast to the atherosclerotic process where typically both internal mammary arteries are disease free, syphilitic vasculitis often involves middle-sized arteries including the internal mammary arteries and may thus preclude their use as coronary bypass conduits. Arterial cannulation for cardiopulmonary bypass using the axillary or femoral arteries rather than the aorta is preferred to reduce the risk of cerebrovascular complications.

Connective tissue disorders and syphilis can be associated with massive hemolysis in the presence of hypothermia as a result of the interaction between cold agglutinins and red blood cells; therefore, normothermic cardioplegia should be used to avoid this possible interaction. Treatment with penicillin before surgery may also carry the risk of Jarisch-Herxheimer reaction that can cause acute aortic inflammation and abrupt closure of the coronary ostia. Neurological involvement is common in syphilitic aortitis. Patients with syphilitic aortitis who have unexplained delirium or neurological deficits should undergo lumbar puncture to rule out neurosyphilis.

CONCLUSIONS

We report a case of syphilitic aortitis suspected by the intraoperative appearance of the aorta. The diagnosis was confirmed by aorta pathology and postoperative serological testing. Given the increase in the number of primary syphilis cases that have occurred during the human immunodeficiency virus epidemic, it is important to remain aware of the clinical and pathological manifestations of tertiary syphilis, including syphilitic aortitis.

REFERENCES