A 37-year-old man presented to our department complaining of severe chest pain triggered by the slightest physical exercise. The symptoms appeared 6 months previously and have gradually worsened. His medical history was unremarkable, and he had no cardiovascular risk factors except smoking. A 6-lead ECG at rest was normal, as was the chest x-ray. The ultrasonography examination revealed severe aortic regurgitation and a moderately enlarged left ventricle with normal systolic function. The treadmill test was positive for myocardial ischemia. He subsequently underwent a catheter examination that diagnosed severe right and left main coronary ostial stenoses along with severe aortic valve incompetence (Figure 1A and 1B). A computed tomographic examination confirmed these findings (Figure 1C and 1D), revealing an ascending aorta with a maximum transverse diameter of 3.5 cm, an irregular intima, and a thickened wall (6 mm) (Figure 1C). He was referred for urgent replacement of the aortic valve and triple total arterial coronary bypass.

At surgery, the ascending aorta had a hyperemic, inflammatory adventitia that adhered firmly to the surrounding structures. Cross-clamping revealed a markedly thickened wall, with extensive longitudinal wrinkling all the way into the aortic root, deforming and narrowing the coronary ostia (Figure 2). The aortic valve leaflets were thickened and retracted, resulting in severe, central incompetence. Aortic valve replacement with a 25-mm mechanical prosthesis and a triple total arterial coronary bypass with the use of the radial and both internal thoracic arteries was performed.

Figure 1. Coronary angiography showing ostial stenosis of the right (A) and left coronary artery (B). C. Computed tomographic scan revealing a thickened aortic wall (6 mm), an irregular intima at the level of the aortic root, and right coronary ostial stenosis. D. Ostial stenosis of the right coronary artery (RCA) and left main coronary artery (LMCA).
arteries was performed. The pathological examination revealed endarteritis obliterans of the vasa vasorum, a chronic inflammatory infiltrate in the medial layer with disruption of the elastic fibers, and a severely thickened intima (Figure 3A and 3B). See the online-only Data Supplement for additional information. A Venereal Disease Research Laboratory test was performed and was intensely positive, as was the Treponema pallidum hemagglutination test. For further confirmation, we attempted to identify treponemal DNA in the pathological sample by means of the polymerase chain reaction. However, this test did not reveal traces of living treponemal spirochetes. The patient had an uneventful recovery and was treated with 2.4 million units of intramuscular benzathine penicillin G for 3 weeks. Subsequent testing remained negative at 1 year. The patient is followed up by periodic computed tomography scans to detect any enlargement of the thoracic aorta.

Discussion

Better sexual education and treatment have contributed to a marked reduction in the incidence of syphilis. However, migration of the workforce in a globalized world, drug use, and sexual promiscuity have created conditions for sexually transmitted diseases to recur.

Untreated syphilis can reach the tertiary phase with cardiovascular and neurological manifestations. The classic clinical symptom is that of an ascending aortic aneurysm that is typically large and saccular. Progression may lead to erosion of the sternum, pseudoaneurysm formation, or outright dissection. Intramural hematoma of the ascending aorta has also been described recently in this context. The pathological hallmark is endarteritis obliterans of the vasa vasorum with a chronic inflammatory infiltrate, ischemic necrosis, and fibrosis of the medial layer. Scarring of the aortic media leads to longitudinal wrinkling of the aortic wall (tree barking); however, extension of the disease into the aortic root with coronary ostial stenosis appears to occur in a minority of cases. Tree barking of the ascending aorta is also seen in systemic lupus erythematosus and other systemic inflammatory diseases. Aortic valve regurgitation results from inflammation, fibrosis, and retraction of the cusps as well as from dilatation of the sinotubular junction. Death is a consequence of aortic wall rupture, chronic heart failure, cerebrovascular accidents, or acute myocardial infarction.

Definitive diagnosis of tertiary syphilis is based on indirect (Venereal Disease Research Laboratory test) or direct (Treponema pallidum hemagglutination test) methods. Identification of living spirochetes by polymerase chain reaction in tissue samples of patients has been described recently; however, the sensitivity of this method appears to be low.

Tertiary syphilis remains a diagnosis to be considered in patients with aortic pathology, especially if it is associated with coronary ostial stenosis with a normal coronary bed.

Disclosures

None.

References

Coronary Ostial Stenosis in a Young Patient
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