Unusual Case of Nonbacterial Thrombotic Endocarditis Attributable to Primary Antiphospholipid Syndrome

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A 54-year-old man was referred for cardiac evaluation before renal transplantation because of chronic renal failure. His medical history was remarkable for diabetes mellitus, hypertension, and 3 episodes of transient ischemic attack (the most recent was 3 years earlier). A transthoracic echocardiogram showed a large, mobile mass on the anterior leaflet of the mitral valve, with trivial mitral regurgitation (Figure 1; Movie I in the online-only Data Supplement). The mass was originally believed to be a fibroelastoma; endocarditis was not considered initially because of the absence of systemic symptoms. Transesophageal echocardiography showed a 1×2-cm mobile, echodense region on the A2 segment of the mitral valve leaflet (Figure 2 and Movie II in the online-only Data Supplement are 2-dimensional images; Figure 3 and Movie III in the online-only Data Supplement are 3-dimensional images). The patient underwent open cardiac surgery for removal of the mass (Figure 4). Histological examination showed nonbacterial thrombotic endocarditis (NBTE). The patient was discharged from the hospital to his home with long-term anticoagulation therapy. To establish the cause of NBTE, he underwent computed tomography of the chest, abdomen, and pelvis, but no obvious underlying malignancy was identified; however, a seroimmunologic blood test was positive for lupus anticoagulant not associated with connective tissue disease. Therefore, the diagnosis of primary antiphospholipid syndrome was made. In retrospect, this likely was the cause of his transient ischemic attacks.

Discussion

We report an unusual patient with NBTE attributable to primary antiphospholipid syndrome who presented with a large, mobile mass on the anterior mitral valve leaflet. In echocardiography, mobile densities on valve structures usually are vegetations from subacute bacterial endocarditis, tumors, or (rarely) thrombi. In the absence of symptoms, benign tumors such as fibroelastomas often are considered. The finding of a large, intracardiac mass caused by NBTE, without known malignancy or connective tissue disease, is rare.

Vegetations caused by NBTE are found in ~1.2% of autopsies, but the reported prevalence varies widely (0.3% to 9.3%) and predominantly involves patients with coexisting malignancy, sepsis, burns, or disseminated intravascular coagulation.1 However, 60% of patients who undergo surgical...
resection for an intracardiac mass caused by NBTE have autoimmune disease. Fibroelastomas generally are round masses with a “shimmering” appearance on echocardiography; masses are generally attached either to the mitral or aortic valves on the aortic or left atrial side. NBTE vegetations are typically small (< 1 cm), are broad-based, and have irregular borders. Although echocardiographic images may suggest a particular cause, no echocardiographic finding can establish the diagnosis for a specific patient with certainty.

Our case underscores the need for thorough coagulation, malignancy, infection, and connective tissue disease evaluation in any patient presenting with an intracardiac mass and a history of transient ischemic attack so that early diagnosis can be made. Generally, treatment aims to control the underlying condition. Because NBTE is associated with platelet-rich fibrin thrombi, it is possible to treat some small, early stage masses with anticoagulation medication. The most effective anticoagulant appears to be unfractionated heparin, although cardiac surgery should be considered in select patients, depending on the risk/benefit ratio, if they have large, mobile masses and a high risk of embolization.

Disclosures

None.

References

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