A 51-year-old man with Marfan syndrome and multiple previous surgical interventions presented with an enlarging aortic root pseudoaneurysm. His past history was significant for type A aortic dissection at age 25 years, treated with composite root replacement with a Bjork-Shiley valved conduit. Revision of the left coronary button was performed 1 year later as a result of pseudoaneurysm formation. At age 48 years, he had documented ventricular tachycardia with an echocardiogram showing inferolateral hypokinesis. His cardiac enzymes were negative, and he received an implantable cardioverter defibrillator (ICD). Although asymptomatic, he was referred to the Marfan Clinic at our institution a few months later. Transthoracic echocardiography showed mobile density in the ascending aortic graft (Figure 1), and computed tomography (CT) angiography showed a large amount of intraluminal thrombus in the ascending aorta graft, as well as coronary button aneurysms (Figure 2). Magnetic resonance imaging of the head showed infarcts in multiple vascular distributions. The patient underwent redo aortic root replacement with a 25-mm CarboMedics composite graft that required a short segment of a saphenous vein interposition graft to the small left coronary button. A large amount of intraluminal thrombus within the ascending aorta was noted at the time of surgery (Figure 3), and histological evaluation of this thrombus revealed Ebstein-Barr virus (EBV)-associated diffuse large B-cell lymphoma (DLBCL). The neoplastic lymphocytes expressed CD20, MUM1, and BCL2 and in situ hybridization studies evaluating for EBV-encoded ribonucleic acid showed diffuse positivity within the neoplastic lymphocytes. There was no evidence of immunodeficiency or systemic involvement of lymphoma with negative bone marrow biopsy and positron emission tomography/CT scan. This was thought to represent virally mediated malignant transformation of lymphoid cells, likely influenced by the local immune response to the presence of the graft material. No systemic therapy was given. He was maintained on aspirin and warfarin.

One year later, the patient presented with chest pain while exercising, and CT angiography showed a focal irregular filling defect at the ostium of the bypass graft to the left main coronary artery causing severe obstruction (Figure 4). The patient underwent fourth-time redo sternotomy and bypass grafting with a left internal mammary artery to the left anterior descending artery; severe adhesions precluded grafting of the circumflex system. Repeat positron emission tomography/CT showed no evidence of malignancy. However, 18 months later, the patient developed atypical chest and back pain, low-grade fevers, chills, night sweats, and a 15-lb unintentional weight loss. He also complained of clumsiness and weakness of the left arm. CT of the head showed a new right middle cerebral artery distribution infarct. CT angiography of the chest showed an aortic root pseudoaneurysm measuring 32 mm in maximum diameter (Figure 5). Transthoracic echocardiography demonstrated mobile echodensities on the aortic prosthesis and ICD leads (Figure 6). Blood cultures and serologies were all negative. Positron emission tomography/CT showed no evidence of lymphoma or inflammation. Hematologic evaluation revealed no evidence of systemic lymphoma, and rheumatologic evaluation revealed no evidence of primary vasculitis. The patient was treated for culture-negative prosthetic valve endocarditis for 4 weeks with broad-spectrum antibiotics. He then underwent fifth-time redo median sternotomy and aortic root replacement with a 25-mm aortic homograft, reimplantation of the right coronary artery with saphenous vein interposition graft, and ICD extraction. Operative findings included edema of the mediastinal tissues and extensive thrombus or soft tissue involving the valve, graft, and ICD leads. Histology again revealed EBV-associated DLBCL (Figure 7). No bacterial or fungal organisms were identified. The postoperative course was uneventful, and he was discharged 10 days later after implantation of a dual-chamber ICD. The patient is currently undergoing systemic chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone because of the recurrent nature of the lymphoma.

This patient’s case was previously reported in the literature along with 2 other cases of DLBCL involving cardiac prostheses or grafts. The authors felt this case should be classified as DLBCL associated with chronic inflammation according to the 2008 World Health Organization classification of lymphomas and hypothesized that the malignant transformation of lymphocytes occurs as a result of the chronic immune response to synthetic material. They also hypothesized that these cases should have a better prognosis than other cases.
of DLBCL with chronic inflammation because of the resectability. To our knowledge, our case is the only reported case of recurrent DLBCL involving a heart valve or graft material. Serological testing performed postoperatively was consistent with previous EBV infection in this patient, with no evidence of active infection. We do not feel that sufficient evidence exists to suggest that establishing the EBV-exposure status of the patient would be helpful in planning the surgical approach. In this case, as in all of the previously reported cases, the diagnosis was made histologically after the operation, thus making the use of the EBV-exposure status in the planning of the operation not feasible. An aortic homograft was implanted in this case because the working diagnosis at the time of operation was endocarditis; however, we speculate that this may also be less likely than prosthetic material to initiate a chronic inflammatory response. Systemic chemotherapy has been initiated given the recurrence after resection alone.

Disclosures
None.

References

Figure 1. Transthoracic echo image from the suprasternal notch shows a large echodensity in the ascending aorta graft with mobile components (arrow). RPA indicates right pulmonary artery.

Figure 2. Oblique coronal reformat from computed tomography angiography shows a large amount of intraluminal thrombus (arrows) in the ascending aorta graft. Coronary button aneurysms are also seen (asterisks). LV indicates left ventricle; RA, right atrium; RPA, right pulmonary artery; and RV, right ventricle.

Figure 3. Intraoperative photo shows a large amount of thrombus (arrow) in the ascending aorta graft, extending up from the prosthetic valve and along the wall of the graft.
Figure 4. Axial computed tomography angiography image showing an irregular filling defect (arrow) at the ostium of the left coronary artery interposition graft causing severe stenosis of the graft. AA indicates ascending aorta; DA, descending aorta; and RVOT, right ventricular outflow tract.

Figure 5. Oblique coronal reformat from computed tomography angiography showing a large pseudoaneurysm (asterisk) extending posteriorly from the ascending aorta graft and causing mass effect on the proximal left anterior descending and circumflex coronary arteries. Small filling defects are seen attached to the undersurface of the aortic prosthesis (arrow). AA indicates ascending aorta; LV, left ventricle; and RA, right atrium.

Figure 6. Still frame from transthoracic echocardiography imaging shows mobile echodensities attached to the aortic prosthesis (arrow) and cardioverter defibrillator lead (arrowhead). LA indicates left atrium; RA, right atrium; and RV, right ventricle.

Figure 7. Histopathology of Epstein-Barr virus–associated diffuse large B-cell lymphoma (original magnification, ×400). A, The resected thrombus exhibiting enlarged and atypical lymphocytes at the surface of the fibrin-rich thrombus (hematoxylin and eosin staining). B, The atypical lymphocytes demonstrate strong reactivity with antibodies directed against CD20 (dark brown cytoplasmic staining). C, In situ hybridization studies using probes for Epstein-Barr virus-encoded ribonucleic acid shows strong reactivity within the neoplastic cells (dark blue nuclear staining).