Aortic pseudoaneurysms associated with Takayasu's arteritis in a 10-year old boy

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PRE-OPERATIVE CARDIAC EVALUATION WAS REQUESTED for a 10-year-old boy scheduled for excisional biopsy of a painful and possibly malignant fibular mass. This had developed 1 month earlier, and he had a 2-month history of general weakness. Right pulmonary arterial stenosis had been diagnosed the previous year.

On physical examination, the patient had good pulses in all extremities, and blood pressure was normal. Auscultation revealed a grade 3 diastolic murmur heard at the left 3rd intercostal space. The Mantoux test provoked a 15-millimetre forearm



Figure 1.

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induration. C-reactive protein was 5.46 mg/dL, and the erythrocytic sedimentation rate was measured at 55 millimetres per hour. Transthoracic echocardiography showed severe aortic regurgitation due to ectatic change of the aortic root, with the diameter of the aortic sinuses measured at 37.3 millimetres, and pseudoaneurysms of the ascending aorta (Fig. 1;





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Figure 3.

arrows). The presence of multiple pseudoaneurysms, with tears identified in the medial aortic wall, were confirmed by 3-dimensional computed tomography (Fig. 2; thick arrows), while a tomographic scan of the whole body using fluorodeoxyglucose positron emission showed hypermetabolic lesions in the left fibula and ascending aorta (Fig. 3; thin arrows). Biopsy of the fibular mass revealed chronic inflammation.

The aortic regurgitation and the pseudoaneurysms were treated by commissuroplasty of the aortic valve, and replacement of half of the ascending aorta using a 20 millimetre Hemashield graft. Pathological findings showed myxoid degeneration, multinucleated giant cells, and formation of abscesses, compatible with Takayasu's arteritis. The patient also received prednisolone 1 mg/kg/day.

At the first follow-up visit to our outpatient department, 1 month after the surgical procedures, C-reactive protein and the rate of erythrocytic sedimentation had normalized. The patient remained asymptomatic when last seen 14 months after surgery, although he had been noted to have steroid-dependent activity of the disease process.