An unusual case of severe aortic valve stenosis in an adult caused by aortic valve inflammatory myofibroblastic tumor

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A 51-year-old male patient reported a history of cardiac palpitations and low fever for 2 weeks. Physical examination showed grade III to IV systolic murmur at the aortic valve

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area. Laboratory examination showed mild anemia (hemoglobin 115 g/L) and increased serum C-reactive protein level (66 mg/L). Echocardiography showed severe aortic valve stenosis with a pressure gradient of 110 mm Hg caused by a tumorlike lesion (Figure 1, A-C). The patient underwent aortic valve replacement. During the operation, a granulomatous tumor approximately $2.0 \times 2.0 \times 2.5$ cm was found with a thickened aortic valve (Figure 1, D). Histocytologic examination revealed an inflammatory myofibroblastic tumor (IMT) with hyperplastic inflammatory spindle cells and pleomorphic myofibroblasts. Immunohistochemical analysis showed it to stain highly positively for smooth muscle actin and also positively for vimentin and desmin.

Cardiac IMT is a very rare disease, and only sporadic reports appear in the literature. Because most cardiac

FIGURE 1. A-C, Echocardiographic studies demonstrating severe aortic valve stenosis with a pressure gradient of 110 mm Hg caused by a tumorlike lesion. D, Intraoperative photograph. LV, Left ventricle; M, myofibroblast tumor; RA, right atrium; RV, right ventricle.

IMTs are found in children, an IMT in an aortic valve causing severe aortic stenosis is extremely rare in the adult. The diagnosis of IMT is not easy; thus it was important that our doctors knew the disease and recommended that the pathologist perform appropriate immunohistochemical analysis. Our patient had traditional IMT symptoms, such as fever and anemia, as well as

palpitations related to valvular disease. At the same time, laboratory examination showed an increased serum C-reactive protein level. The diagnosis was later supported by echocardiography, pathologic examination, and immunohistochemical analysis. This was a classic case of IMT in the aortic valve causing severe aortic valve stenosis.