Cardiac involvement of Wegner granulomatosis: It's rare but not unheard of

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Granulomatosis with polyangiitis (GPA; ie, Wegner granulomatosis) is a antineutrophil cytoplasmic antibodies-associated systemic vasculitis that predominantly affects the small vessels with focal necrotizing, granulomatous lesions.^{1,2} The incidence of GPA is very rare; it usually appears in 40-year-old patients and most commonly involves the otolaryngological tract (92%), the renal (glomerulonephritis; 77%), and the pulmonary system (85%).¹ In contrast, direct cardiac lesions beyond an unspecific pericardial effusion are reported in the literature in approximately 6% to 44% of patients.² Lacoste and colleagues³ summarized in a contemporary literature review 20 cases of systemic antineutrophil cytoplasmic antibodies-associated GPA with cardiac involvement. The most frequent manifestations were conduction disorders (atrioventricular block), coronary artery disease, and valvular disorders, especially due to aortic (57%) and mitral valve regurgitation (14%).^{3,4} Although resolution of the valvular manifestations might be achieved by immunosuppressive therapy in some cases, valve replacement was necessary in most patients to resolve heart valve incompetence.

In this issue of *The Journal of Thoracic and Cardiovascular Surgery*, Abe and coworkers⁵ report their experience with 4 additional cases of granulomatosis with polyangiitis (GPA)-associated cardiac disorders requiring cardiac surgery. Clinical symptoms and positive c-antineutrophil cytoplasmic antibodies confirmed the GPA diagnosis accompanied by meticulous preoperative imaging using computed tomography and cardiac magnetic resonance imaging that revealed tumor mass in the aortic root with extension to the fibrous body of the mitral as well as tricuspid valve. As nicely depicted in the illustrations of the report, the authors proceeded to surgery with extensive and radical resection of the mass followed by complex

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Central Message

This report presents clinical decision pathways with respect to diagnosis, symptoms, and surgical resection strategies in patients with cardiac manifestation of granulomatosis with polyangiitis.

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reconstruction of the mitral annulus and left atrium after valve replacement. The pathological findings of the specimen confirmed a chronic necrotizing granuloma highly suggestive for GPA.

The authors can be congratulated for their aggressive approach, which seems justified on the basis of the excellent surgical results and uneventful postoperative course of all patients. Importantly, this case series underscores that treatment of GPA-associated cardiac lesions is feasible and can be safely performed. As a prerequisite for successful surgery, preoperative imaging (computed tomography, magnetic resonance imaging) seems to be imperative to identify the exact anatomical position and feasibility of surgery, and to exclude other malignant cardiac mass entities (lymphomas, sarcoma, etc), that might be a contraindication for cardiac surgery and/or more amendable to conservative treatment strategies. Finally, although surgery seems to be a viable option for GPA-associated cardiac lesions the long-term results of this chronic autoimmune disease including late cardiac recurrence despite surgery and optimal immunosuppressive therapy is still largely unknown. Nonetheless, we cardiac surgeons need to be aware of this rare but treatable cardiac disease.

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