An ugly nose and a Nazi father: Bad premise for a cardiac rheumatic disorder

Massimiliano Foschi, MD,1 Fabrizio Costantino Tancredi, MD,2 and Michele Di Mauro, MD, PhD3

The rheumatic disorder reported on by Abe and coworkers1 in this issue of the Journal has long been labeled as Wegner granulomatosis, ever since Friedrich Wegener,2 a Nazi pathologist working in Poland,3 reported 3 cases in 1937, describing the pathologic disorders as vasculitis. Actually, this disease had first been reported by Wegner’s friend, a Berlin pathologist named Heinz Klinger,4 who in 1932 described a case of destructive sinusitis, lung abscesses, and uremia, characterized by angiitis and granulomas, in a report entitled, “Borderline Forms of Periarteritis Nodosa.” Today, this rheumatic disorder is known as granulomatosis with polyangiitis (GPA), and one of the most typical sign is a saddle nose deformity caused by a perforated septum, along with crusting around the nose, stuffiness, and runny and bleeding nose,5—in few words, an ugly nose!

GPA is a form of vasculitis of small and medium vessels associated with anti–neutrophil cytoplasmic antibodies. Although it typically involves the upper and lower airways, lungs, and kidneys, cardiac involvement is estimated in as many as 25% of cases in the general population and as many as 44% among patients with severe renal involvement.6

The most common cardiac manifestations are pericarditis and coronary arteritis, which are usually medically treated. Although valve involvement is not actually uncommon (57.1% for aortic valve and 14.3% for mitral valve), as previously reported by Abe and coworkers,1,7 there is little evidence of surgical treatment for these cases. The strength of this case series is therefore its use of surgery as a treatment, with excellent results, in cases in which GPA severely affected the fibrous heart structure, resulting in multiple valvulopathies, but we need a worldwide registry.

Our future challenge in the field of rare heart diseases that can be surgically addressed, such as GPA, could and should be to establish worldwide registries to evaluate the results of surgery on larger cohorts.

References