Multi-modality cardiac imaging for granulomatosis with polyangiitis

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In this issue of the Journal of Nuclear Cardiology, Al-Mehisen et al. present a case of a 21-year-old woman with dyspnea, atypical chest pain, and intermittent palpitations associated with epistaxis, stuffy nose and frontal headaches found to have granulomatosis with polyangiitis (GPA).¹ Echocardiography showed mild left ventricular systolic dysfunction with a small apical aneurysm in addition to reduced global longitudinal strain. Cardiac magnetic resonance imaging (CMR) demonstrated myocardial edema in the anterior and lateral wall and late gadolinium enhancement (LGE) in the septum suggestive of fibrosis. The patient was suspected of having sarcoid but later work-up, including positive c-antineutrophil cytoplasmic antibody (c-ANCA), was suggestive of GPA. With no response to anti-inflammatory treatment, the patient underwent further imaging with 18F-fluoro-2-deoxyglucose (FDG) positron emission tomography (PET) which revealed matched perfusion-metabolism defect in the septum and apex consistent with scarring, and evidence of cardiac inflammation in other areas. She was placed on the heart transplant waiting list but was lost to follow-up and presented four years later with acute heart failure and developed cardiac arrest due to ventricular fibrillation and expired.

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GPA is one form of the ANCA-associated vasculitides which also include microscopic polyangiitis and eosinophilic granulomatosis with polyangiitis.² GPA has been long known as Wegner's granulomatosis since the 1950s after Dr. Friedrich Wegener who characterized the disease in the 1930s although it was described earlier by Klinger. However, in the 2000s when the Lancet was going to write an article celebrating Dr. Wegner, it uncovered his secret Nazi past.³ He had joined the Nazi party in 1932, even before Hitler took power in Germany and rose to high military rank with some suggestions that he participated, or at least knew of experiments on concentration camp inmates. This led the American College of Chest Physicians to rescind its designation of Wegener as a master clinician, which it awarded him in 1989 the year before he died.⁴ It further led to the formal change in the name of the disease from Wegner's granulomatosis to GPA.^{5,6}

Although the diagnosis of GPA should be established by tissue biopsy, multi-modality imaging can provide important contributions to the management of patients with GPA.⁷ As demonstrated in this case, the use of echocardiography, CMR and PET may help identify and differentiate areas of cardiac fibrosis and inflammation with resultant implications for treatment. The use of FDG PET for active cardiac inflammation is well defined for cardiac sarcoid,^{8,9} but further studies are needed to better characterize its role in vasculitides such as GPA.

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