Regarding "Waxing and Waning Presentation of Isolated Cardiac Sarcoidosis on Sequential ^{18}F-FDG PET Exams"

TO THE EDITOR: I read with interest the article by Ostwani et al. (*1*) and, for 3 reasons, am concerned that the patient does not have cardiac sarcoidosis (CS). First, because of the lack of a positive biopsy result, the patient does not meet guideline criteria for the diagnosis of CS (*2*). Second, corticosteroid-refractory sarcoidosis is considered very rare (*3*). Third, accumulating data suggest that there may not be a pathophysiologic entity of truly isolated CS. It is clear that there are many patients with manifest CS who have no clinically apparent disease in other organs—that is, who have what can be termed *clinically* isolated CS. However, sarcoidosis is, by definition and biology, a systemic disease. Hence, a key starting point to understand isolated CS is to agree on a standardized definition. The 2017 version of the Japanese CS guidelines tackled, for the first time, the definition of and criteria for the diagnosis for isolated CS (*4*). They included the following 3 criteria: no clinical findings characteristic of sarcoidosis are observed in any organs other than the heart; ^{67}Ga scintigraphy or ^{18}F-FDG PET reveals no abnormal tracer accumulation in any organs other than the heart; and a chest CT scan reveals no shadowing along the lymphatic tracts in the lungs or no hilar and mediastinal lymphadenopathy (minor axis $>$ 10 mm).

Using a similar definition, my group found *imaging*-isolated CS in only 1 in 31 patients (*5*). However, other data suggest that even these apparent isolated cases are unlikely to be truly isolated. Petek et al. investigated 10 patients with presentations and cardiac imaging consistent with the Japanese definition of isolated CS. Four of these 10 had granulomas on bronchial biopsy (*6*). Hence, these data suggest that there is a small subset of patients who at the moment of ^{18}F-FDG PET imaging have PET-detectable inflammation only in their heart. However, it also follows that additional or interval investigation will likely reveal extracardiac disease.

This debate is more than just semantics, as the overdiagnosis of "imaging-isolated CS" can, as in this case (*1*), lead to unnecessary immunosuppression or a missed alternative diagnosis.

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