The Curious Case of Subcutaneous Sarcoidosis

Meagan Herda, MD
VA Portland Health Care System

Introduction
Sarcoidosis is a multisystem granulomatous disease that can affect any combination of organ systems. Although the vast majority of patients are diagnosed due to pulmonary symptoms or findings of hilar lymphadenopathy, as many as 25% of patients with sarcoidosis have skin findings. However, because of the high variability in their presentation, the initial diagnosis of sarcoidosis is rarely made based on dermatologic manifestations.

Case Presentation
A 67 year old Caucasian female with obesity, Type II diabetes mellitus, osteoarthritis, gout and hypothyroidism presented to clinic with subcutaneous nodules on her bilateral upper extremities. Two years prior to presentation, the patient had a rash on her lower extremities. Biopsies at that time revealed both a neutrophilic dermatisis as well as a lymphocytic infiltrate. The differential remained broad, but she was given a diagnosis of Sweet Syndrome caused by NSAID use. The offending agent was discontinued and her skin lesions resolved. However, in the months prior to presentation, the patient had struggled with fatigue, depression and unintentional weight loss for which previous workup had been unrevealing. Although she also had been experiencing these nodules for several months, she had not previously mentioned them to her provider as she thought it was simply a recurrence of Sweet Syndrome.

Physical Exam
Vitals: BP 133/83 | Pulse 86 | Temp 98.4 °F | RR 16 | SpO2 94%
Weight 254 lbs - 82)
Gen: obese female in no acute distress
HEENT: no oral lesions, no cervical adenopathy
CV: regular rate and rhythm, no murmurs, rubs or gallops
CHEST: clear to auscultation without cracks or wheezes
EXT: numerous subcutaneous nodules over dorsum of bilateral forearms and hands ranging from 1-5 cm in diameter, some of which are tender and slightly erythematous. Several raised superficial nodules involving the tattoo on her right forearm.

Laboratory Data
TSH: 2.34 U/mL (ref 0.27-4.2)
Hgb 11.5 g/dL MCV 80 f (baseline Hgb 13 w/ MCV 90)
Creatinine 1.3 mg/dL (baseline 0.7)
Calcium: 11.9 mg/dL (ref 8.4-10.1)
PTH: 6 pg/mL, PTHrp negative
Vitamin D 25.23 ng/mL (ref 30-100)
Vitamin D 1,25- 98 pg/mL (ref 18-72)
ACE: 79 U/L (ref 14-82)
Rheumatoid Factor, c-ANCA, p-ANCA negative, ANA: 1:1280

Clinical Course
The patient was diagnosed with panniculitis. Initial rheumatologic studies were unrevealing except for an elevated ANA. Workup of her hypercalcemia revealed low PTH and PTHrp, elevated 1,25-dihydroxyvitamin D and borderline elevated angiotensin converting enzyme. Imaging of her forearms was obtained which revealed subcutaneous edema and inflammation (Figures 1 and 2). She was referred to surgery for biopsy which revealed abundant noncaseating granulomas. With this biopsy result and the aforementioned studies, the patient was diagnosed with subcutaneous sarcoidosis. Subsequent CT scan of the chest, abdomen and pelvis revealed diffuse lymphadenopathy as well as several lung nodules (Figure 3) consistent with systemic sarcoidosis. Pulmonary function testing showed mild restrictive lung disease. Given her hypercalcemia and systemic symptoms, she was treated with a prolonged taper of high dose steroids (prednisone 60mg daily) and hydroxychloroquine. After only a month of treatment, the patient reported a subjective improvement in her energy, her calcium had declined to 9.5 and her vitamin D 1, 25 decreased to 19. At her most recent visit, she was found to have a new first degree AV block (Figure 4) for which an echocardiogram is pending. An ophthalmologic exam was obtained and was normal. In retrospect, the patient’s initial diagnosis of Sweet Syndrome was likely Sarcoidosis. However, due to the nebulous pathology and scarcity of subcutaneous sarcoidosis, this diagnosis was overlooked.

Discussion and Teaching Points
- It is important to recognize atypical presentations of illnesses.
- We recognize pulmonary, cardiac and ocular manifestations of sarcoidosis, but overlook dermatologic presentation.
- Raised nodules involving old scars or tattoos should make you think of cutaneous sarcoidosis.
- Patients with subcutaneous sarcoidosis are at high risk for systemic disease and should undergo further testing.
- It is important to question diagnoses made by other providers as this likely delayed the patient’s request for a skin exam and delayed her diagnosis and treatment of sarcoidosis.

References