

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 dermatosis 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 -> 241 lbs

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 crackles 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 fl (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

Imaging

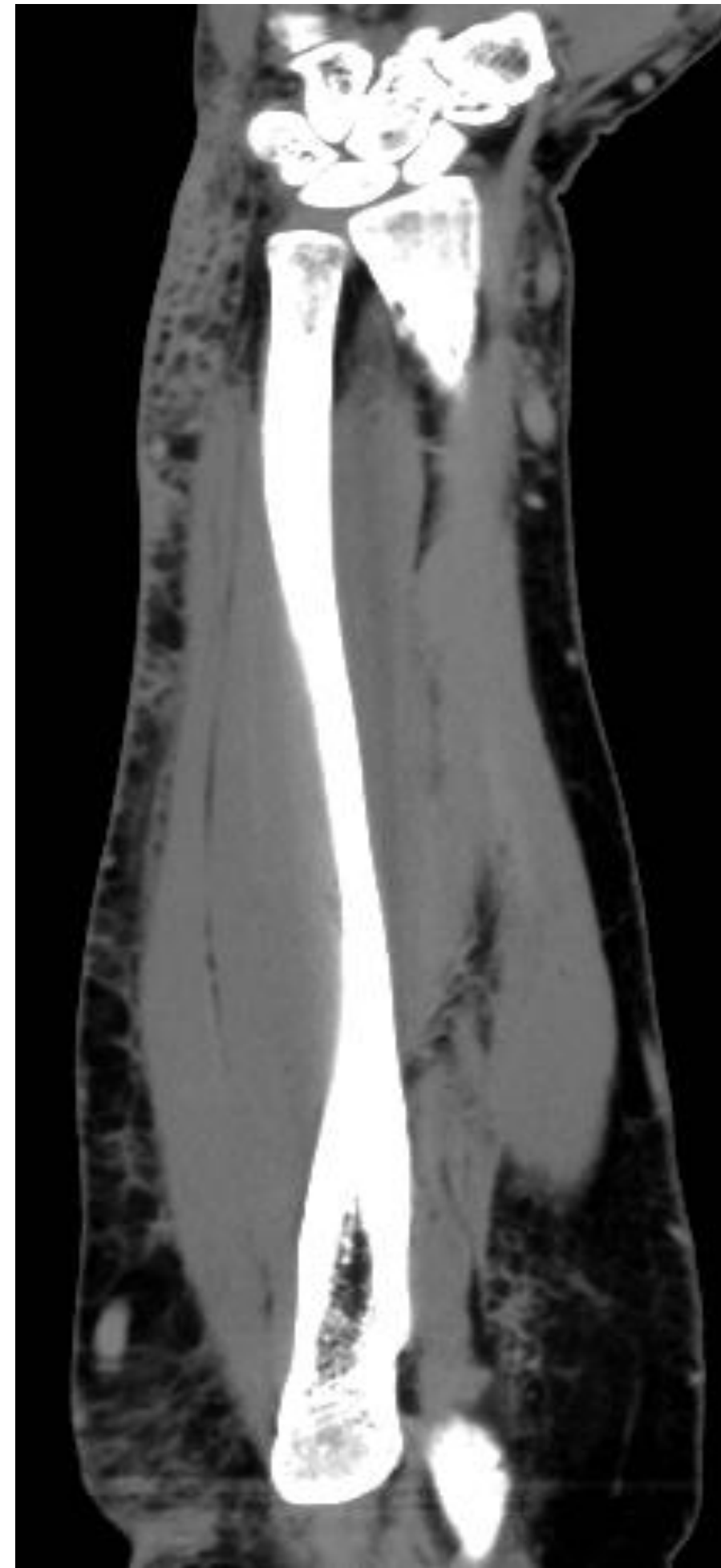


Figure 1. CT of the forearm revealing subcutaneous edema and inflammation.

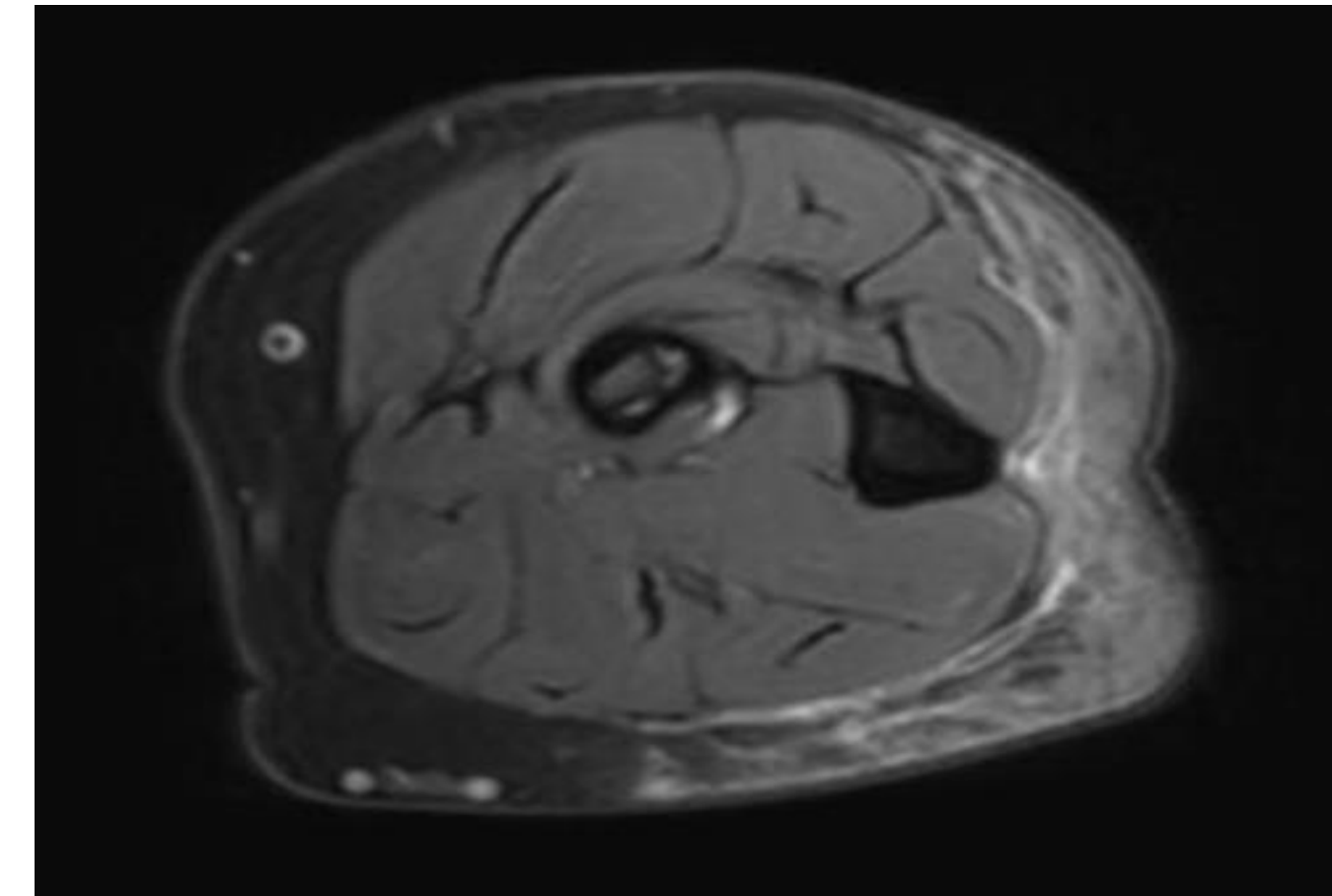


Figure 2. T2 enhanced MRI of the forearm revealing subcutaneous edema and inflammation.



Figure 3. CT of the chest demonstrating hilar lymphadenopathy.

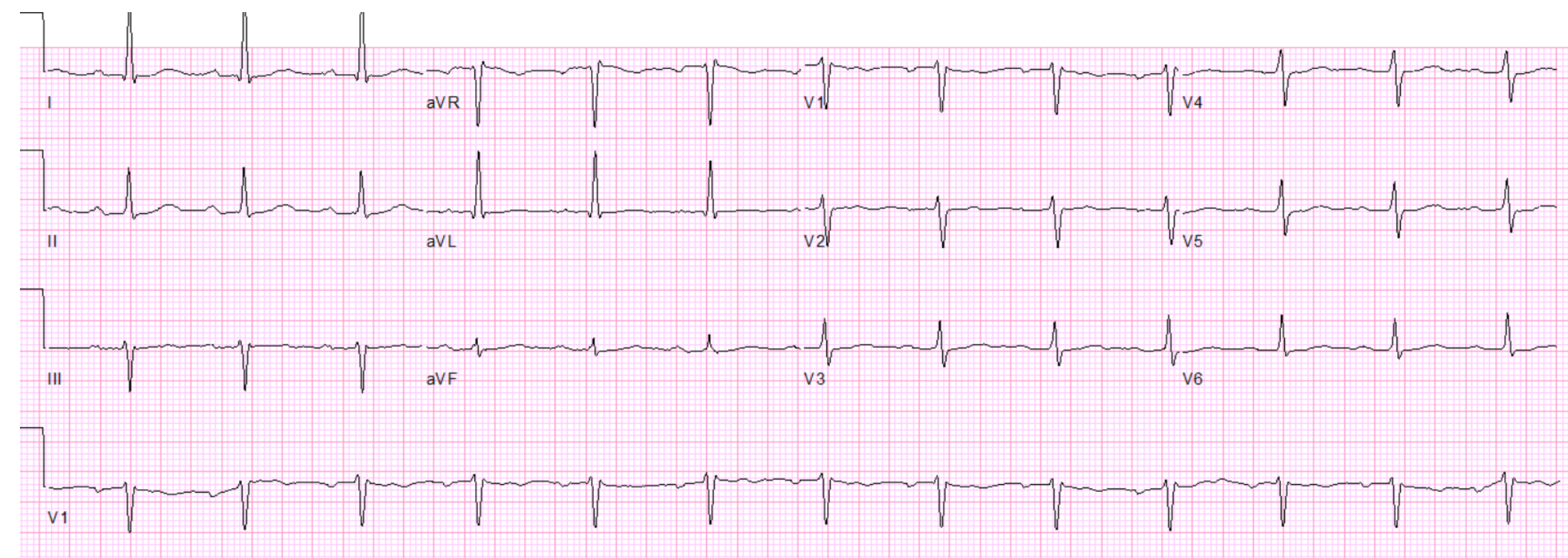


Figure 4. EKG with new 1st degree AV block concerning for cardiac involvement.

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

1. Iannuzzi MC, Rybicki BA, Teirstein AS. "Sarcoidosis". *New England Journal of Medicine*. 357 (2007): 2153-2165.
2. Ahmed I, Harshad SR. "Subcutaneous sarcoidosis: Is it a specific subset of cutaneous sarcoidosis frequently associated with systemic disease?" *Journal of the American Academy of Dermatology*. 54.01 (2006): 55-60.
3. Torres LK, Faiz SA. "Tattoos and Sarcoidosis". *New England Journal of Medicine*. 370.e34 (2014): 24.