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CLINICAL VIGNETTE

Sarcoidosis: A Case Study

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Case Report

A 56-year-old female presented to her primary care physician with complaints of dyspnea on exertion and increasing cough. She reported no fevers, chills, night sweats, or hemoptysis, however, did relate an approximate 20-pound weight loss over 6 weeks prior to presentation. She reported no history of sick contacts or recent travel.

Her past medical history was significant for hypertension, dyslipidemia, osteopenia, and idiopathic thrombocytopenic purpura. The patient does relate a history of sarcoidosis diagnosed 20 years prior to presentation where she had an asymptomatic left supraclavicular lymph node biopsied and confirmed to be sarcoidosis. She did not receive any medical therapy at that time and her adenopathy resolved spontaneously. Her medications included lisinopril, simvastatin, actonel, and temazepam at the time of evaluation. She reported no known malignancies in her family history and her social history was positive for lifelong smoking but negative for alcohol or any illicit drug use.

Physical Examination:

The patient's general exam was within normal limits except for multiple, scattered erythematous to violaceous and tender skin nodules on her bilateral extremities. Her lungs were clear to auscultation and she had no palpable adenopathy.

Initial Laboratory Values:

White Blood Cell Count $5.07 \times 10^3/uL$, Hemoglobin 12.3 g/dL, Hematocrit 38.2%, Platelet 59,000. Total Bilirubin 0.5 mg/dL, AST 30 U/L, ALT 21 U/L, Alkaline Phosphatase 68 U/L. Angiotensin Converting Enzyme 41 U/L. ANA and Rheumatoid factor were negative. Her chemistry panel revealed no abnormalities.

Pulmonary Function Tests:

FVC 3L (88% predicted), FEV1 2.54L (94% predicted), Total Lung Capacity 4.64L (86%) predicted. The rest of her spirometry tests were normal.

Imaging:

Chest X-Ray taken demonstrated evidence of micronodular interstitial densities throughout bilateral lung fields with apical fibronodular granulomatous changes. Follow-up Chest CT revealed innumerable centrilobular micronodules throughout the lungs predominantly in the upper lobes. Mediastinal and hilar adenopathy was also noted.

Pathology:

Punch biopsy of a right pretibial skin lesion showed a well-formed sarcoidal type of granuloma with septal thickening and fibrosis. PAS stain was negative for fungal organisms and there was no evidence of vasculitis or malignancy. Final reading was consistent with sarcoidosis.

Treatment Course:

Patient was initiated on prednisone 60 mg per day for 6 weeks and then tapered down 5 mg every 4 weeks. After 4 months of therapy, platelet counts normalized to 181,000. The patient's spirometry results had also improved: FVC 3.25L (94% predicted), FEV1 2.56L (94% predicted), Total Lung Capacity 5.4L (100% predicted). Serial chest radiographs also demonstrated a corresponding improvement. The patient reported no further dyspnea on exertion or any constitutional symptoms.

Discussion:

The incidence of sarcoidosis is slightly higher in women than in men, with an incidence of 6.3 and 5.9 cases per 100,000

person-years, respectively. The lifetime risk of sarcoidosis for Caucasians in the United States is estimated at 0.85 percent compared with 2.4 percent in U.S. African-Americans. Although sarcoidosis can appear at any age, a bimodal age pattern is seen, which peaks between ages 25-35 and 45-65 years¹.

Sarcoidosis is characterized by noncaseating epithelioid granulomas that can affect virtually any organ system. The diagnosis of sarcoidosis requires the biopsy finding of noncaseating granulomas not caused by any other etiology. The disease most commonly involves the lungs (see Table 1 – sarcoid staging system in lungs) but can also affect the liver, skin, heart, nervous, and endocrine systems. The exact etiology of sarcoidosis is unknown. Evidence points to a possible genetic component, infectious etiology, or potential exposure to environmental agents². Infectious organisms such as mycobacteria and *Borrelia burgdorferi* have been implicated as potential causes of sarcoidosis. Environmental exposure to beryllium, aluminum, and zirconium can result in a granulomatous response similar to that of sarcoidosis. Current theories suggest that the disease develops in genetically susceptible hosts who are exposed to certain antigens that trigger an overwhelming inflammatory immune response leading to granulomatous reactions³.

Because sarcoidosis can involve any organ system, the clinical presentation is variable. The onset can be insidious and findings may be discovered only on routine chest radiographs. Cutaneous involvement is seen in 25% of patients with sarcoidosis and it usually accompanies systemic involvement but may be the only site of involvement. Table 2 lists some of the most common clinical signs.

Oral corticosteroids are the main treatment for pulmonary sarcoidosis. In a review of corticosteroids for pulmonary sarcoidosis, treatment with oral steroids for 6 to 24 months improved chest x-ray findings compared with placebo. Patients with interstitial lung disease (stages II and III) had benefits in both global scores and chest radiographs. Data show that no treatment is necessary for patients with stage I disease. Table 3 lists the many available modalities of treatment that exist for sarcoidosis⁴.

Patients should be evaluated after one to three months for response. Those who fail treatment after initial corticosteroid therapy usually will not respond to a more protracted course of treatment. In responders, the prednisone dosage should be tapered to 5 to 10 mg per day or to an every-other day regimen, and therapy should continue for up to a year. There is no consensus guidance on treatment beyond two years. Patients must be monitored after cessation of treatment for possible relapse as some patients will require long-term low-dose therapy to prevent recurrent disease. The risk of osteoporosis must also be accounted for in patients taking prolonged systemic corticosteroids. Although few studies have addressed this issue, the bisphosphonate alendronate has been shown to prevent osteoporosis in patients with sarcoidosis⁵.

The course of the disease is variable, but spontaneous remission occurs in 50% of patients, while most improve with corticosteroid treatment. About 10-30% of patients have chronic or progressive disease. The mortality rate is approximately 1-6%. (1) In the United States, mortality is most commonly due to respiratory failure from pulmonary involvement, cardiac involvement, or neurosarcoidosis. Complications of therapy are additional causes of morbidity and mortality.

Table 1- Stages of Sarcoidosis

Stage	Chest X-Ray Characteristics
0	Normal
1	Bilateral hilar adenopathy
2	Bilateral hilar adenopathy and parenchymal infiltrates
3	Parenchymal infiltrates without bilateral hilar adenopathy
4	Signs of fibrosis

Table 2 – Clinical Signs/Symptoms

- **General:** 1/3 of patients have constitutional symptoms, such as fever, fatigue, and weight loss.
- **Pulmonary system:** The lungs are affected in most patients. Symptoms occur in up to 1/2 of patients and most commonly include dyspnea, dry cough, and chest pain.
- **Lymphatic system:** 1/3 of patients with sarcoidosis have palpable lymphadenopathy.
- **Ocular involvement:** Anterior uveitis is the most common finding, which may be associated with fever and parotid swelling (also called uveoparotid fever). Chronic uveitis, most commonly occurring in African Americans, can lead to adhesions, glaucoma, and cataract formation.
- **Neurosarcoidosis:** Involvement of the nervous system occurs in fewer than 10% of patients, however, it can be lethal. The disease can affect any part of the nervous system. Seventh cranial nerve palsy is the most frequent finding. The pituitary gland and the hypothalamus may be involved.
- **Myocardial involvement:** Clinically apparent cardiac involvement occurs in 5% of patients in the United States.

Table 3 - Treatment

- Oral corticosteroids are usually the treatment of choice for patients with neurologic, cardiac, or ocular involvement not responding to topical corticosteroids, hypercalcemia, and symptomatic stage II/III pulmonary disease. The usual dose is 30-40 mg of prednisone daily for 2-3 months, with a gradual taper over 1 year to 10-20 mg every other day. Patients with severe disease may need prednisone doses up to 1 mg/kg/d.
- Limited cutaneous involvement may be treated with topical or intralesional corticosteroids. Intralesional injections of 2-10 mg/mL of triamcinolone acetonide can be used at monthly. (6) If intralesional corticosteroids ineffective, other standard therapies including systemic corticosteroids, methotrexate, and antimalarials (hydroxychloroquine and chloroquine) can be used. (7)
- Infliximab is a promising option for those with recalcitrant or disfiguring disease. (8)
- Other agents that have been used to treat sarcoidosis include cyclosporine, and chlorambucil. (9)

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