CLINICAL VIGNETTE

Sarcoid Arthropathy

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Case Report

A 46-year-old caucasian male presents to the rheumatology for chronic bilateral knee pain. His past medical history is significant for pulmonary sarcoidosis that was diagnosed 10 years ago. CT imaging at the time showed enlarged mediastinal and hilar lymph nodes, as well as diffuse abdominopelvic adenopathy and mild splenomegaly. A lymph node biopsy confirmed the diagnosis of sarcoidosis. He had no history of erythema nodosum or cardiac involvement but developed episodes of uveitis thought to be related to his sarcoidosis with flares occurring approximately every 9 months. He was treated with various courses of prednisone tapers on and off for his pulmonary or ocular flares that were effective treating his symptoms. He had no other significant past medical history and was on no medications on presentation.

Over the last 6 years, he developed intermittent knee pain and swelling, with the right knee more affected than the left. There was no history of trauma. He had tried physical therapy and ibuprofen with minimal benefit but did note improvement in his pain during steroid courses given for his sarcoidosis. Knee x-rays were normal. Labs for ANA, rheumatoid factor, CCP antibodies and HLA B27 were all normal. ACE levels also normal. MRI of the right knee showed a large knee joint effusion with severe synovitis consistent with sarcoid arthritis.

He was started on daily prednisone 10mg with resolution of his knee pain and swelling. Methotrexate was added and titrated up to 20mg a week as a steroid sparing agent. The prednisone was slowly tapered over 6 months without recurrence of the arthritis. His pulmonary symptoms and uveitis flares also improved on the methotrexate minimizing the need for courses of systemic steroids.

Discussion

Sarcoidosis is a systemic inflammatory disorder of unknown etiology characterized by a noncaseating granuloma reaction that predominantly affects the lungs and intrathoracic lymph nodes,[1] but may also involve other organs.[2] Sarcoidosis is found in all races and ethnic groups worldwide with the highest incidence in Scandinavian countries. In the United States, there is a higher incidence of sarcoidosis in the African American population when compared to Caucasians.[3] Generally, sarcoidosis is more common in women and mostly occurs between 20-40 years of age, although a second peak has been reported in women over the age of 50.[2]

Sarcoidosis can present with various clinical manifestations. The disease presents most often with bilateral hilar lymphadenopathy, pulmonary infiltrates, uveitis and skin lesions.[2] Skin lesions can include erythema nodosum, lupus pernio, inflammatory papules and ulcers. Lofgren's syndrome, a triad of bilateral hilar adenopathy, erythema nodosum and arthritis has long been described as a unique presentation of sarcoidosis.[1] In sarcoidosis, the lungs are the most commonly involved organ followed by skin, eyes, heart, liver and the musculoskeletal system.[3] This patient's sarcoidosis involved lung, spleen, ocular and joint, as clinical manifestations.

Joint involvement or sarcoid arthropathy is reported in 6-35% of patients with sarcoidosis.[4] Two major patterns of joint involvement have been defined: acute and chronic forms. The most common is the acute form which can be the first symptom of sarcoidosis[2] and precede the diagnosis of sarcoidosis by an average of 3 weeks.[1]

Oligoarthritis of the bilateral ankles was the most common presentation of acute sarcoid arthritis. Hilar adenopathy and erythema nodosum as seen in Lofgren's syndrome were the most common extra-articular features associated with acute sarcoid arthritis.[5] The prognosis of acute sarcoid arthritis is generally favorable as the arthritis can resolve within 6 weeks in the majority of patients.[2]

Chronic arthritis is observed more commonly in patients with diffuse organ involvement[3] and is associated more frequently with ocular involvement[4] as seen with this patient. In a large series of patients with chronic sarcoid arthritis, upper limb joints (predominantly distal) and knee involvement were the most common joints affected.[5] In severe cases, the chronic arthritis may cause a Jaccoud's type deformative arthropathy and erosive changes on radiography[3] although this is rare. ANA and rheumatoid factor serologies may be positive as well, mimicking other rheumatologic conditions.[2] Synovial biopsies show presence of noncaseating granulomas.[3]

Spontaneous remission is reported in the majority of patients with acute sarcoid arthritis. In some cases, non-steroidal anti-inflammatory drugs or low dose oral corticosteroids may be required for pain management. Colchicine and hydroxychloroquine may also be used in treating Lofgren's syndrome and erythema nodosum. For chronic sarcoid arthritis, immunosuppressive drugs such as methotrexate, leflunomide, azathioprine

and anti-TNF agents can be used for treatment.[3] This patient responded well to methotrexate treatment reducing his flares and overall need for systemic steroids.

In conclusion, arthritis can be an extrapulmonary manifestation of sarcoidosis and chronic forms of sarcoid arthritis, although less common, can occur as presented in this patient. Long term immuno-suppressive therapy may be required in these cases for better disease management.

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