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Case Report
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# Lofgren's Syndrome: A Rare Sarcoidosis Presentation in a Rural Hospital

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## Abstract

Lofgren's syndrome is the most common presentation of acute sarcoid arthritis, it presents as a triad of hilar adenopathy, acute polyarthritis and erythema nodosum. Acute polyarthritis generally presents in <2 months and classically involves ankles bilaterally. Erythema nodosum is also classically present upon initial symptoms. Chest radiograph is needed to make a diagnosis of sarcoidosis and for confirmation of hilar adenopathy. Patient is a 43 year old man who was found to have bilateral ankle and left wrist swelling and pain, later found to have erythema nodosum and hilar lymphadenopathy. Initially started on ibuprofen with partial relief, and then started on oral prednisone with rapid symptomatic relief within 2 months of initiation and resolution of symptoms. This case highlights a typical course and workup of Lofgren's syndrome and evidence guided treatment.

**Keywords: Lofgren's syndrome; Hilar adenopathy; Polyarthritis; Sarcoidosis**

## Introduction

Sarcoidosis is a disease of unknown etiology, characterized by non-caseating granulomas throughout multiple body systems, most commonly involving the lungs (>90% cases) often manifesting as hilar adenopathy [1]. Other body systems can be affected, with cutaneous, joint and ocular being the most often involved [2]. About 25% of patients have cutaneous involvement, which is usually the first or early symptom of sarcoidosis [3]. About 10% of patients will develop joint symptoms, which can manifest as either acute of chronic arthritis [4]. Lofgren's syndrome is the most common presentation of acute sarcoid arthritis (5% to 10% of sarcoid cases), it presents as a triad of hilar adenopathy, acute polyarthritis and erythema nodosum (though not all components are necessary to make a diagnosis) [5,6]. Acute polyarthritis usually involves ankles (>90%) and other large joints of the lower extremity, less commonly it involves wrists and smaller joints of hand [7,8]. The joint involvement is usually symmetrical and oligoarticular and manifests more often in males [9]. Erythema Nodosum (EN) are erythematous and tender nodules along shins bilaterally [10]. EN's pathogenesis is delayed-type hypersensitivity, though specific antigens and immune complexes have not been identified, it is thought to involve immune complex deposition in venules and subcutaneous fat of shins that elicits an immune response8. EN can be triggered by many different processes including: infections, drugs, malignancies and various inflammatory diseases (including sarcoidosis) and is usually self-limited, often resolving within a few weeks after onset [10]. EN manifests more commonly in women than men with Lofgren's syndrome [9]. There are no standardized confirmatory tests for this syndrome, though chest radiograph is needed to make a diagnosis of sarcoidosis and for confirmation of hilar adenopathy [5].

Lofgren's syndrome is self-limited, erythema nodosum, hilar adenopathy and acute polyarthritis usually resolve within a few weeks to months, however polyarthropathy can last for up to 2 years [4-6]. Lofgren's syndrome has been linked to human leukocyte antigen (HLA) DR3 and DQ2 alleles which seem to correlate with more persistent polyarthritis. HLA-DQB1*0201 and DRB1*03 alleles seem correlated to less severe disease course [12]. Treatment involves symptomatic control with NSAIDs/colchicine or oral glucocorticoids until symptoms resolve, if disease is resistant to these therapies, hydroxychloroquine, methotrexate or infliximab can be used [5,4].

## Case Presentation

Patient is a 43-year-old male who presented initially to Emergency Department (ED) with left ankle swelling and bruising as well as left leg pain and swelling in anterior portion, these symptoms had been ongoing for a few weeks prior; a referral was made to rheumatology for arthritis as well

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as started on ibuprofen for symptomatic management. Patient noted to never have any arthritis symptoms up until this point in time, and these symptoms started abruptly without obvious trigger. Upon initial visit to rheumatology clinic, was found to have symmetrical polyarthritis: Bilateral ankle swelling and pain, worse in left ankle, as well as left wrist pain and swelling, EN over lower extremities as well as coughing and shortness of breath that had been ongoing for a few weeks. Patient had improvement of pain and swelling of joints on ibuprofen. ESR and CRP were elevated at 74 and 6.7 respectively. Chest X-ray was obtained to assess shortness of breath and coughing, was found to have bilateral hilar and right paratracheal lymphadenopathy. Diagnosis of Lofgren's syndrome was made and patient was started on steroid taper, initially started at 60 mg with a taper that was weaned off in 6 months. Symptoms resolved within 2 months of steroid initiation, including lymphadenopathy, hilar lympadenopathy and polyarthritis with ESR and CRP returning to 5 and <0.5 within this time period as well.

## Discussion

Lofgren's syndrome is an uncommon presentation of sarcoidosis occurring in only about 5% to 10% of sarcoid patients. It is, however, important to recognize as it is the most common form of acute sarcoid arthritis and prompt treatment can prevent unnecessary prolonged discomfort for patients. Although no formal standardized testing has been established, the triad of hilar adenopathy, erythema nodosum and acute polyarthritis has classically been used for clinical diagnosis.

In the case described above, our patient presents with the classical triad of Lofgren's syndrome. Acute polyarthritis, especially bilaterally in the ankles (a classical presentation in males) as well as additional oligoarthritis in left wrist developed. It is important to note that the patient had no prior history of arthritis and that symptoms started acutely, and had progressed quickly within the span of about one month. This correlates to the acute arthritis picture that is classic for Lofgren's syndrome. EN and hilar lymphadenopathy were also noted at the same time as acute polyarthritis. While the patient happen to develop dyspnea and cough and obtained a chest x-ray, suspicions should have been raised given the presentation of EN and acute bilateral ankle polyarthritis of Lofgren's syndrome since two of the triad had already presented themselves. A chest X-ray is a critical component of diagnosing Lofgren's syndrome because it is a quick and easy way to assess for presence of pulmonary sarcoidosis, thus establishing both a diagnosis of sarcoidosis and by proxy, Lofgren's syndrome.

Although diagnosis of Lofgren's syndrome wasn't made at the time of ED visit, patient was started on ibuprofen, with partial symptomatic relief prior to initial rheumatology visit. But ibuprofen by itself wasn't adequate for symptom control, thus oral prednisone was started and provided rapid symptomatic relief within 2 months of initiation. Of note, due to the efficacy and quick resolution of symptoms, no further interventions were initiated for this patient, however should symptoms have persisted with oral glucocorticoids, further interventions with methotrexate or hydroxychloroquine could have been initiated, and infliximab beyond that. Further workup to qualify disease process with HLA alleles could also have been performed.

This case was a classical presentation of Lofgren's syndrome that had quick resolution of their disease with relatively minimal treatment. It is fortunate that a rheumatology referral was made from the ED as many cases of Lofgren's syndrome are often missed or delayed in being diagnosed as arthritis is such a common symptom. With the EN, this should prompt suspicion for sarcoid arthritis and Lofgren's syndrome, and confirmation with chest X-ray to determine presence of hilar lymphadenopathy. This case highlights the typical presentation and proper workup process of classic Lofgren's syndrome as well as evidence based treatments.

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