Case Report

Lofgren's syndrome, a presentation of Sarcoidosis: A Case report

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Abstract

Sarcoidosis, a granulomatous disorder of unknown cause, is an autoimmune multisystem condition that primarily impacts adults between the ages of 20 and 40. While the condition can affect nearly any organ in the body, it typically manifests as pulmonary infiltrates, skin lesions, eye issues, and joint problems. Lofgren's syndrome, characterized by acute arthritis, erythema nodosum, and bilateral hilar lymphadenopathy, is an unusual initial presentation of sarcoidosis. . We describe here a newly diagnosed case of sarcoidosis that presented as Lofgren's syndrome. Acute sarcoid arthritis should be kept as one of differential diagnosis for patient presenting with acute arthritis and skin lesions. Chest X Ray should be considered to rule out bilateral hilar lymphadenopathy. Early suspicion and recognition of typical features are crucial for making a diagnosis.

Introduction

Sarcoidosis is a granulomatous disease of unknown etiology common in young adults and middle age people.[1] It affects almost every organ mainly intrathoracic as bilateral hilar adenopathy and diffuse lung micronodules while extrathoracic manifestations as skin lesions, uveitis, peripheral lymphadenopathy and peripheral arthritis.[2] Acute onset arthritis is characterized by symmetrical arthritis while chronic arthritis is characterized by persistent oligoarthritis or polyarthritis along with arthralgia.[3] Lofgren's syndrome, seen in 5 -10% of sarcoidosis is characterized by acute arthritis, erythema nodosum and hilar lymphadenopathy.[4] We present a 33 year old woman who presented with Lofgren's syndrome.

Case Presentation

A 33-year-old married female presented to our medical OPD from Chitwan, Nepal without known comorbidities with a history of bilateral ankle swelling for the last 2 months which was gradual and progressive associated with mild pain that aggravated on walking. She was treated earlier with various medications including NSAIDs with only transient relief of signs and symptoms. She had no history of low back pain, small joint pain, or joint stiffness prior. She also gave a history of tender swelling on limbs on and off that used to resolve spontaneously with dark pigmentation. There was no history of cough or shortness of breath. On examination, her vitals were - Blood pressure 110/70 mm of Hg, Pulse 80/min, SpO2 97% in room air, and afebrile. On local examination, she had bilateral ankle swelling without restriction in movement and mild tenderness. Rest systemic examination was normal on examination.

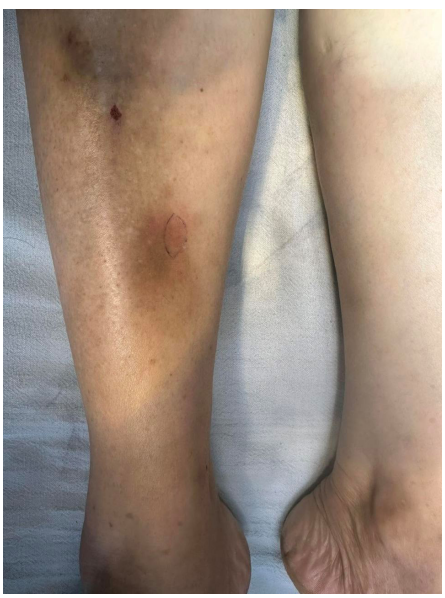

Figure 1. Erythema Nodosum

Her laboratory examination revealed CBC, RFT LFT within normal limits, S. Albumin 3.7gm/L, S. Calcium 9.0 mg/dl, ANA, and Anti CCP Negative. However, CRP was positive at 14.2 mg/L. Her other lab findings showed an Angiotensin-converting enzyme level of 60 U/L (N-12-68 U/L), 24 hours Urinary Calcium level of 329.3 mg/24 hours. (N – 42.0 – 353 mg/24 hours). Her Chest x-ray

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showed findings suggestive of bilateral hilar adenopathy so she was suggested for HRCT chest which revealed mediastinal and bilateral hilar lymphadenopathy with few perilymphatic micro nodules in bilateral lungs and sub pleural reticulations in bilateral lower lobes suggesting features of Sarcoidosis (Stage II). Her X-ray of pelvis/SI Joints was normal.

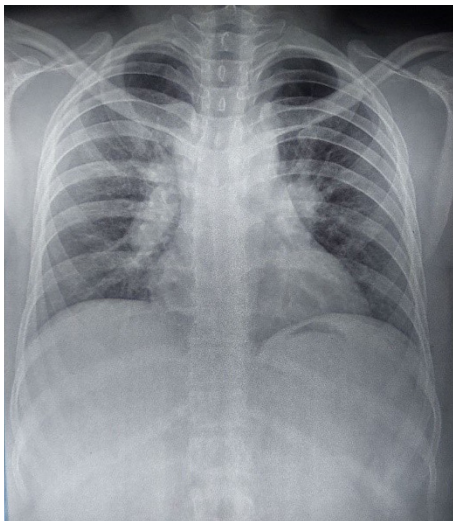

Figure 2. Chest X ray showing Hilar adneopathy

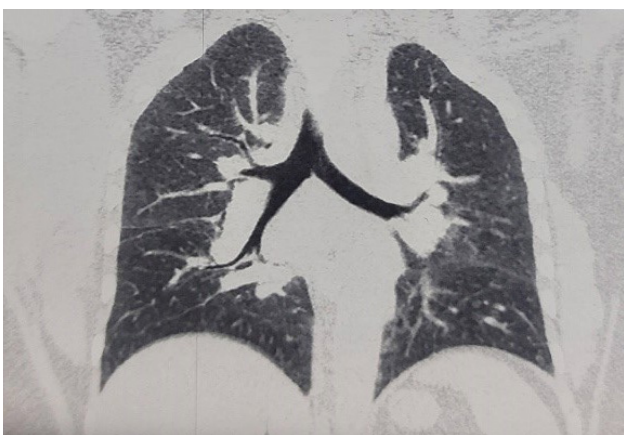

Figure 3. CT showing Hilar adenopathy

With the diagnosis of Sarcoidosis, she was initiated on Tab Prednisolone 40mg with tapering dosages. After a week of therapy, she had relief of ankle swelling. Currently, she is on a tapering dose of steroids and has been initiated on Methotrexate as an immunosuppressant.

Discussion

Sarcoidosis is a multisystem disorder of unknown etiology. In Nepal, the prevalence of sarcoidosis is not well known and Lofgren's is rarely reported[5] It is common in the Brahmins community in Nepal and should be in differential diagnosis of young patients with mediastinal or hilar lymphadenopathy with or without infiltrates.[5] The disease affects almost every organ in the body but commonly present with bilateral hilar lymphadenopathy, pulmonary infiltrates, cutaneous and joint lesions. Joint involvement also known as sarcoid arthropathy is seen in 6 – 35% of patients[3,4] Bone and joint manifestations in sarcoidosis may include Lofgren's syndrome, which presents as acute arthropathy, as well as chronic disease featuring granulomatous synovitis and tenosynovitis. Other manifestations may involve Jaccoud's type arthropathy, dactylitis, and non-articular bone sarcoidosis.[6]

Lofgren's syndrome is a specific acute clinical presentation of systemic sarcoidosis and has excellent prognosis with more than 90% chance of spontaneous remission within 2 years.[7] Erythema nodosum occurs more frequently in women than in men who exhibit Lofgren's syndrome. Approximately 15% of patients with the initial manifestation of Lofgren's syndrome have elevated ACE levels in the peripheral blood and experience persistent arthritis.[4] However elevation of ACE differs in various population as it is seen in 30.7% of Asian patients while 54.1% in European population.[8] Predisposing factors and prognosis of Lofgren's syndrome may be associated with race, sex, cytokines, and gene polymorphism. There are also reports of HLA-DRB1*03 is associated with Lofgren's syndrome and its prognosis.[8]

Conclusions

Sarcoidosis is a common condition. We reported here a case of a 33-year-old female patient with Lofgren's syndrome, exhibiting bilateral hilar adenopathy, acute onset Erythema Nodosum, multiple arthritis, and arthralgia. She achieved remission rapidly with corticosteroid administration. Lofgren's syndrome is rarely reported especially in Nepal so further studies and clinical experience are needed.

Conflict Of Interest

None

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