CASE REPORT

ARTHRALGIA IN A YOUNG PATIENT: PRESENTATION OF LOFGREN SYNDROME

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Lofgren syndrome an acute form of sarcoidosis constitutes erythema nodosum, bilateral hilar adenopathy and arthralgia or arthritis. Here we present a case of a 28 years old young male patient who is a shopkeeper and farmer by profession admitted to inpatient department with chief complaints of bilateral painful nodules on his shins, low grade fever and pain multiple joints on both sides of the body. Suspicion of the Lofgren syndrome was made upon initial evaluation and patient was admitted to inpatient care facility for necessary comprehensive workup. Radiological findings were consistent with bilateral hilar lymphadenopathy and patient was diagnosed with Lofgren syndrome. Patient was started on non-steroidal inflammatory drugs (NSAIDs) with close observation for improvement in response to treatment. After one weak of treatment in hospital, patient was discharged home when his symptoms started to resolve.

Keywords: Sarcoidosis; Erythema nodosum; Arthralgia

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INTRODUCTION

Sarcoidosis is a granulomatous disease that involves multiple systems.[1] It can involve muscles, tendons, ligaments, and joints.[2] Lungs are the most commonly involved organs (90%). Among other commonly affected organs are Skin, eyes, liver and lymph nodes. At the time of presentation 14% of the patients have joint involvement, and then at follow up the frequency goes up to 38%. Sarcoidosis most frequently occurs in women than in men. Worldwide incidence is reported to be 16.5–19 /100,000.[3–5]

Lofgren syndrome has been reported in patients with origin from North Europe, Ireland and japan. Lofgren syndrome is characterised by erythema nodosum, bilateral hilar lymphadenopathy, polyarthritis and/or arthralgia; is an acute form of sarcoidosis and can be its initial presentation.[5] It is common amongst Caucasians; frequently reported in young women hailing from Nordic countries and Ireland.[6] Prognosis is good with most patients going into complete remission in first 2 years.[7]

CASE REPORT

A 28 years old male patient admitted to inpatient care with painful swellings on both shins, low grade fever and swellings of wrist, knee and ankle joints bilaterally from last two weeks. Fever was low grade, intermittent, with rigors and chills. He also had a dry mouth and nocturia. Patient suffered Bell's Palsy 2 months back which has resolved by now. He was shopkeeper and a farmer by profession and has no history of recent trauma or travel. Family history is significant for hypertension in both parents. On examination, temp was 100°F, Blood pressure 130/85, pulse 92 beats per minute and respiratory rate of 18 per minute. A dry beefy tongue was noted; however, no lymphadenopathy was noted on examination. There was a painful, erythematous, blanching rash on shins with pitting oedema, suggestive of erythema nodosum as examined and confirmed by senior dermatologist, shown in fig 1. Further examination revealed mild stiffness of the knee, ankle and wrist joints symmetrically. On chest auscultation breath sounds were decreased in bases of both lungs. Rest of the systemic examination was unremarkable.

Laboratory studies showed a raised ESR of 45 mm/1st hour, peripheral smear showed anisocytosis, poikilocytosis and raised eosinophils with a value 11% (white blood cells 8100/microlitre), INR 1.7, LDH 262 U/L, ALP 141U/L, normal ALT and total bilirubin. Serum electrolytes ware within normal range, Anti-CCP, rheumatoid factor, ANA were negative, CXR showed bilateral hilar opacities as shown in figure-2, ECG was unremarkable.
CT chest revealed bilateral hilar lymphadenopathy. Centriacinar nodules were seen in the right lower lobe, middle lobe and left upper lobe, there was also peri-bronchi vascular soft tissue thickening around the right lower lobe bronchus, shown in figure-3 and 4. Taking into account all these features patient was diagnosed with Lofgren syndrome. CT scan of the chest reported by Senior radiologists confirmed the diagnosis. Patient was started on Naproxen 250 mg twice a day per oral and other symptomatic treatment. Follow up was scheduled in 6 weeks' time. On follow up patient showed significant improvement from his previous presentation. No new episodes of fever occurred after the discharge from hospital, erythema nodosum was resolved and there was mild pain in bilateral knee joints. On the successive follow up 2 months later, joints pain was also subsided and patient had no complaints.

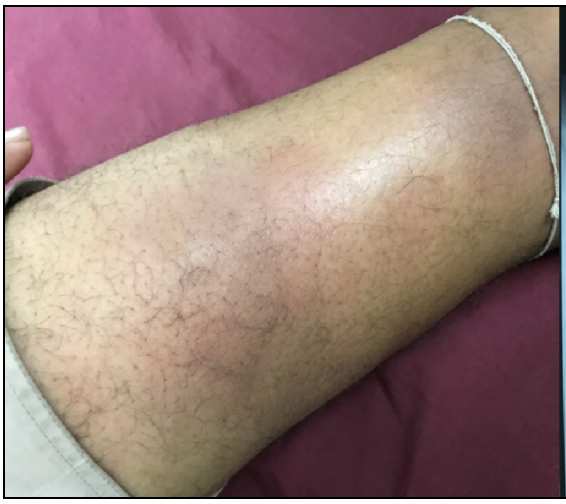

Figure-1: Erythema Nodosum

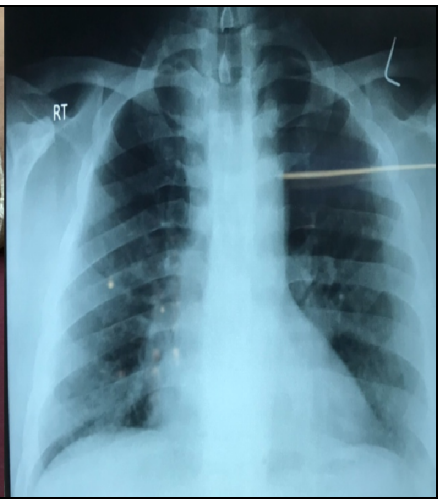

Figure-2: Bilateral Hilar opacities on CXR

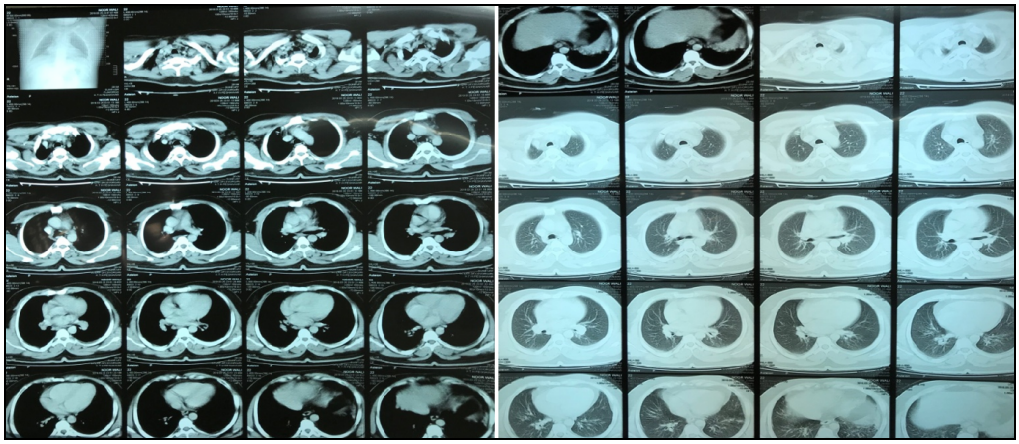

Figure-3,4: Bilateral Hilar lymphadenopathy, peri bronchi vascular soft tissue thickening and centriacinar nodules

DISCUSSION

Lofgren syndrome is quite rare in our setup but it should be considered one of differentials in patients who present with painful red nodules on shins consistent with erythema nodosum. General supportive measures, observation and NSAIDs prove effective in majority of cases and patients carry a favourable outcome. In our case based on history and initial examination differential diagnosis of sarcoidosis and rheumatoid arthritis was established, however, CT scan findings turned out to be helpful and diagnosis of Lofgren syndrome was made an acute form of sarcoidosis which can be its initial presentation as well.

Arthritis in sarcoidosis is frequently symmetrical and greater than 90% of the patients have ankles joint involvement. In 15 to 60% of patients knee and small joints of hands and feet, wrists, and elbows are also involved.[5] A type of sarcoidosis first described in 1953 by Sven Lofgren is Lofgren syndrome.[8] Bilateral hilar lymphadenoapthy, Erythema nodosum (EN) and arthritis or arthralgia are its characteristic features.[9] Some other symptoms include fever in 38% of patients, cough in 13% while hepatomegaly and raised calcium in 6%and 2% respectively. Only 1% had hypertrophy of salivary gland.[9]

In a study consisted of 106 patients with confirmed Erythema nodosum was noted in 22% of cases were found to have Lofgren syndrome or Sarcoidosis. Viral upper respiratory tract infections make up 20%, Group A beta haemolytic streptococci 7%, Tuberculosis 5%, Drugs 3%, Inflammatory bowel disease and malignancy are other causes of erythema nodosum. One third of the cases are idiopathic, i.e., 8%. Therefore, Lofgren syndrome even though rare should be considered while evaluating a patient with Erythema nodosum.[10]

Various Collagen diseases can present with EN and arthralgia. Lofgren syndrome should be kept as a differential here as well. This includes Rheumatoid arthritis (RA).[5] Our patient was also initially suspected to have RA but it was eventually ruled out after necessary evaluation.

Diagnosis of acute sarcoidosis can pose a challenge to clinicians and can be missed as variants of this disorder can distort the clinical picture.[5] For acute sarcoidosis the diagnostic specificity for the characteristic symptoms of EN, bilateral hilar lymphadenopathy and arthritis is 95%.[11] In Lofgren syndrome there is often absence of uveitis and angiotensin converting enzyme (ACE) elevation. These are normally present in sarcoidosis, thus making diagnoses challenging.[5] ACE enzymes are nonspecific and are elevated in about half of patient with EN but their levels can be used to assess resolution in those whom it's initially increased. For evaluation of the hilar lymphadenoapthy a computed tomography (CT) of the chest should be done in patients.[9] As in our case CT scan of chest was consistent with sarcoidosis. To rule out other causes of bilateral hilar adenopathy and erythema nodosum (EN), biopsy is usually done but it's not necessary for the confirmation of the diagnosis. Non-caseating granulomas can be seen on lymph node histopathology.[9]

Mean duration of Lofgren syndrome is several weeks to several months. It is considered a self-limiting disease. But episodes of recurrent sarcoidosis were reported in 6% patients after 2 and up to 20 years. After 2 years of onset 8% of patients still had symptoms.[8]

NSAIDs accompanied by bed rest is the mainstay of treatment in most of the cases. Steroids is indicated in case of in case of severe cases such as severe arthritis, symptomatic hypercalcemia and severe skin lesions. [9] In our case patient responded to NSAIDs alone and showed significant improvement and eventually resolution of all symptoms.

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