Case Report

Lofgren syndrome: Acute Variant of Sarcoidosis, A case report

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Abstract:

Lofgren's syndrome is an acute form of sarcoidosis that is characterized by erythema nodosum (EN), bilateral hilar lymphadenopathy (BHL) accompanied by arthritis or arthralgia. We are reporting a 32 year old male with Lofgren's syndrome and then the literature is reviewed. Keywords: lofgren's syndrome, sarcoidosis

Introduction:

Sarcoidosis is an enigmatic chronic multisystem inflammatory condition of obscure etiology, primarily affecting young adults aged between 20 and 40 years.[1,2] The lungs are the most frequently affected organ (>90%), but the skin, eyes, liver, and lymph nodes are commonly involved.[3,4]. The modern history of sarcoidosis goes back to 1899, when the pioneering Norwegian dermatologist Caesar Boeck coined the term to describe skin nodules characterized by compact, sharply defined foci of "epithelioid cells with large pale nuclei and also a few giant cells." Thinking this resembled sarcoma, he called the condition "multiple benign sarcoid of the skin.[5] There are two eponymous syndromes associated with sarcoidosis. Heerfordt syndrome is the symptom triad of - uveitis, parotid gland enlargement and cranial nerve paresis and was described by Danishophthalmologist C. F. Heerfordt in 1909.[6] Lofgren's syndrome is a form of acute-type sarcoidosis that is characterized by erythema nodosum (EN), bilateral hilar lymphadenopathy (BHL) accompanied by arthritis or arthralgia.[7]. It was first recognized by Sven Lofgren in 1953 in a series of 113 patients." Sarcoidosis affects people of all racial and ethnic groups and occurs at all ages, with the incidence peaking at 20 to 39 year.[10] The highest annual incidence of sarcoidosis has been observed in northern European countries (5 to 40 cases per 100,000 people).[11]. The adjusted annual incidence among black Americans is roughly three times that among white Americans.[10]

In Bangladesh, there is not much statistical data regarding the prevalence or pattern of sarcoidosis. Because of rarity of this disease and inexperience of some cytopathologists, many cases clinically consistent with sarcoidosis are falsely labeled as tuberculosis. We believe that sarcoidosis is more prevalent in Bangladesh than currently known. We are reporting an adult male with Lofgren's syndrome, an acute variant of sarcoidosis, affecting skin, musculoskeletal, ocular and respiratory systems.

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Case Report:

32 years old male presented to us with the complaints of pain in ankles, knees, wrists and elbows, redness and watering from both eyes and vague chest discomfort for 4 weeks. His joint pain was severe enough to his daily activities. He did not have fever cough, night sweats, weight loss, breathlessness, syncope, headaches, nausea, vomiting, abdominal pain, diarrhea, dysuria, and skin rash. He did not give any past history of tuberculosis or recent contact with patients of tuberculosis. Treatment with NSAID's did not help. On examination he was not anemic, lymph nodes were not palpable, and salivary glands were not enlarged. Erythema nodosum was present in both shins. His ankle, knee, wrist and elbow joints were tender on palpation. He had mild redness in eyes Examination of respiratory, gastroinstinal CVS and nervous system were unremarkable. His complete blood count was normal, ESR was 40. RA test, antinuclear antibody was negative, Slit lamp examination of the eyes revealed bilateral anterior uveitis, but his visual acuity was noted to be normal in both eyes. CXR showed bilateral hilar lymphadenopathy (Fig.-1). USG of

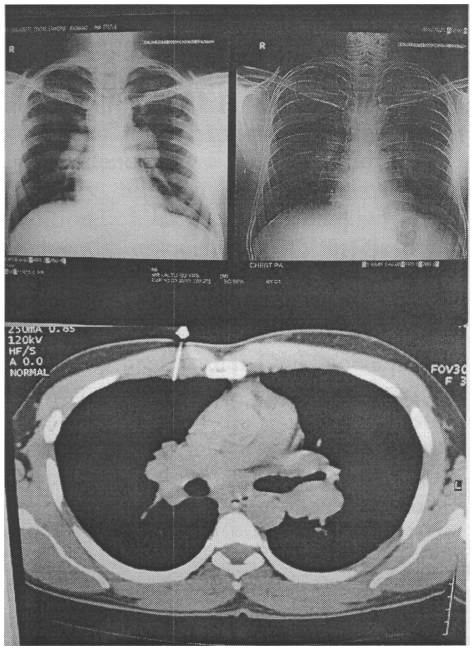

Fig. 1 and 2: CXR and CT scan chest showing bilateral hilar lymphadenopathy

whole abdomen was normal. Mantoux test showed no induration. CT scan of chest demonstrated bilateral hilar lymphadenopathy (Fig. 2),
pulmonary parenchyma was normal.

CT guided FNAC of hilar lymph node was reported as: non-caseating granulomatous inflammation compatible with sarcoidosis (Fig 3, 4). Serum calcium was 9.2 mg/dL. Routine urine examination, liver and renal functions and ECG were normal.

This patient was diagnosed as a case of Lofgren's syndrome based on his classical clinical picture: BHL, EN, and arthritis and supported by presence of non-caseating granuloma on FNAC of hilar lymph node. Prednisolone was started at a dose of 40 mg/day; patient became asymptomatic after 15 days. Prednisolone was started at a dose of 40 mg/day; patient became asymptomatic after 15 days. Prednisolone was being tapered. Bilateral hilar lymphadenopathy regressed after 1 month of steroid treatment. He was Symptomless when last seen.

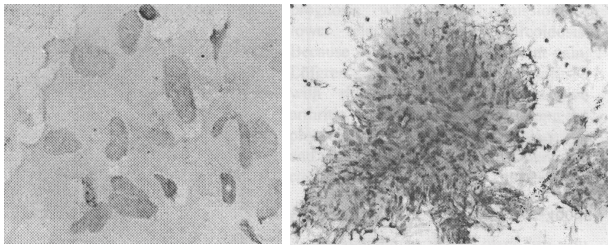

Figure 3 and 4: Showing non- caseating granuloma

Discussion:

The diagnosis of sarcoidosis is commonly established based on clinical and radiological findings that are supported by histological findings. The clinical signs and symptoms are nonspecific and include fatigue, general malaise, weight loss, and less commonly, fever. The characteristic radiological findings associated with sarcoidosis have been well described and the findings include bilateral hilar lymphadenopathy and parenchymal abnormalities.[12] In contrast to other causes of hilar lymphadenopathy, hilar lymphadenopathy in sarcoidosis is typically bilateral and symmetric. In addition, hilar lymphadenopathy is frequently associated with mediastinal lymph node enlargement as depicted on CT scans, especially including the right paratracheal and subaortic nodes. However, mediastinal lymphadenopathy without hilar involvement is rare.[13]. Because of this characteristic distribution, intrathoracic lymphadenopathy typically resembles the shape of the Greek letter lambda (lambda sign).[14]

Some authors believe that histological proof of non-caseating granuloma is necessary to confirm the diagnosis of sarcoidosis.[15] Others noted that presence of BHL along with other classical features of sarcoidosis

has a positive predictive value of 99.95% for diagnosis of sarcoidosis, so routine biopsy poses unnecessary risk to the patient, therefore, not advised.[16]

Up to 35% cases of sarcoidosis are Lofgren's syndrome.[17] . According to Mana et al, who reported a series of 186 patients with Lofgren's syndrome, clinical features of Lofgren's syndrome included arthralgia (68%), fever (38%), cough or dyspnea (13%), granulomatous skin lesion (13%), arthritis (13%), hepatomegaly (6%), ocular symptoms (5%), peripheral lymphadenopathy (4%), splenomegaly (2%), hypercalcemia (2%)[7]. In this series, angiotensin converting enzyme was elevated in about 50% of patients.[7]

Most patients with Lofgren's syndrome have complete resolution of their symptoms, and this syndrome represents a benign acute presentation of sarcoidosis with a good prognosis. In the series of Lofgren, 102 patients (91.9%) had complete resolution of hilar lymphadenopathy and infiltrates, if present, by 2 years.[9] In the series of Maria et al, only 8% of patients (n=11) had active disease by 2 years of diagnosis.[7] In the series of Visser et al, all patients with acute sarcoid arthritis went into clinical remission within 3 months.[18]

Conclusion:

Although L6fgren's syndrome is considered as an acute form of sarcoidosis it has distinct clinical and prognostic features. It has an excellent prognosis, with higher remission or lower relapse rate than sarcoidosis. Because of presentation with joint pain, Lofgren's syndrome might be confused with rheumatoid, viral or reactive arthritis. Increased awareness among the physicians is paramount in the diagnosis and treatment of this rare but potentially curable disease.

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