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Case Report

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Sarcoidosis: A Case Report of an Often-Misdiagnosed Clinical Entity from Bangladesh

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Abstract

A chronic granulomatous disease of unknown etiology without any definite diagnostic criteria with an incidence of 1-1,00,000 among Asians affects multiple systems of our body and is often misdiagnosed owing to its elusive nature and resemblance with other granulomatous disease , especially tuberculosis (TB) in our subcontinent .A 38 years old diabetic woman came with the complaints of low grade fever, night sweats, cough, weight loss, migratory joint pain involving multiple small joints of hands without any morning stiffness and painful skin lesions which resolved spontaneously. Investigation showed Hb 11.5 g/dl with normocytic normochromic anaemia, increased sedimentation, increased CRP, insignificant MT, RA and ANA serology, Sputum for AFB didn't reveal any Mycobacteria. CXR revealed Bilateral Hilar Lymphadenopathy (BHL) and HRCT lungs showed bilateral hilar, mediastinal lymphadenopathy and pulmonary inflammatory lesions. Angiotensin Converting Enzyme (ACE)was 163 U/L which was higher than the normal. She was diagnosed as a case of sarcoidosis and was treated in line with oral prednisolone 40mg/day which was gradually tapered over a period of 5 months. Methotrexate 15mg/day once a week was supplemented with folic acid every consecutive day.Vaccination against H.influenzae and S.pneumoniae were advised and on subsequent follow ups she has been improving .The mortality rate is around 5% however, early suspicion, diagnosis and treatment can be life saving for the patient.

Keywords: Sarcoidosis, Misdiagnosed, Lymphadenopathy, Granulomatous, Elusive

1. Introduction

Sarcoidosis is a multisystem chronic granulomatous disorder of unknown etiology which is mostly a disease of exclusion and goes on to affect the lungs, lymphoid system, skin, eyes, joints, kidney, heart, exocrine glands and the central nervous system [1,2]. While it affects people of all ages, gender and ethnicities but manifestations show a variable range owing to the genetic and environmental factors [2,3]. However, the highest incidence is evidenced among African-Americans which is [7-35/1,00,000] and the lowest among the Asians [1-3/1,00,000] depicting a female and male ratio 2:1 mostly prevalent in non-smokers hailing from rural area [1-3]. Despite having no concrete evidence of the exact etiology there is believed to be a strong association with genetic factors, environmental triggers, infections and autoimmunity. It is often believed to have associations with Mycobacteria, Mycoplasma, Leptospira, Chlamydia and Borrelia [1,2]. Clinical manifestations range from patients being asymptomatic for years to presenting with non-specific manifestations such as nausea, fatigue, fever, loss of weight and occasional night sweats.

Pulmonary symptoms comprise of cough, wheeze, dyspnoea, chest pain, obstructive sleep apnea, hilar and mediastinal lymphadenopathy, the cutaneous variety presents with papules, plaques, subcutaneous nodules, erythema nodusum, scarring, non-scarring alopecia, hyperkeratosis, dystrophy and onycholysis. Renal manifestations range from Glomerular Interstitial Nephritis,

nephrolithiasis, nephrocalcinosis to chronic kidney disease. Ocular features range from uveitis, conjunctivitis, scleritis, episcleritis to even blindness.Cranial neuropathy involving facial and optic nerve are the most common feature of neurosarcoidosis. Diabetes insipidus, adrenal and pituitary failure, galactorrhoea, amenorrhoea are the endocrine presentations. It can go on to present with features of mononeuritis multiplex, GBS, polyneuropathy and polyradiculopathy. The musculoskeletal symptoms range from acute arthritis, arthropathy, osteoporosis, osteopenia to arthralgia. There may be involvement of salivary glands most common being parotid. Lymph nodes mostly involved include cervical, epitrochlear and inguinal.GI manifestations range from vague features of nausea, vomiting and diarrhoea to gross impairment of liver function, hepatosplenomegaly, cholestasis and portal hypertension [1,2,4,5]. It is mostly a disease of exclusion since it has a large number of mimics. As reported by Marc A. Judson in advancement in diagnosis and treatment of sarcoidosis the diagnosis can often be made on clinical grounds without biopsy if all the typical clinical features are present [4]. However, the lack of typical features calls for biopsy, exclusion of other granulomatous disease and proof of involvement of another organ as represented by Figure 1.

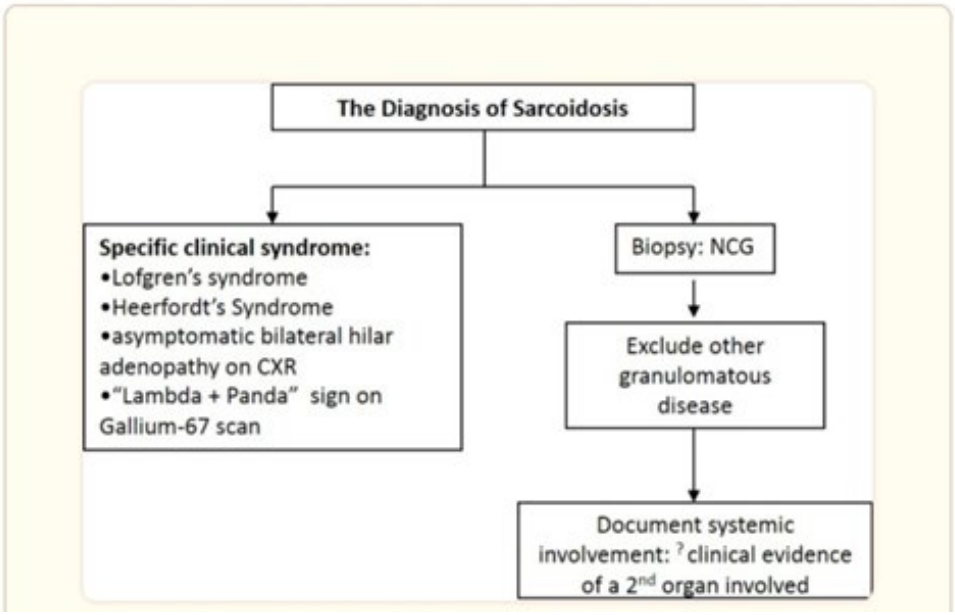

Figure1: Flowchart for the Diagnosis of Sarcoidosis as reported by Marc A. Judson [4].

The non-specific lab tests include CBC, CRP, ESR, and ACE. CXR and CT scan of lung is helpful in diagnosis and radiological classification of sarcoidosis [2,4,5].

Radiological Staging of Sarcoidosis:
Stage 1: Bilateral hilar lymphadenopathy
Stage 2: Bilateral hilar lympadenopathy and reticular opacities
Stage 3: Reticular opacities with shrinking hilar nodes
Stage 4: Reticular opacities with fibrosis [1].

Histopathology reveals non-caseating granulomas although one biopsy isn't confirmative, it does add to the diagnosis in case typical features aren't evident [1,4]. Glucocorticoids remain the main stay of treatment most commonly used of them being prednisolone which is tapered slowly over a period of months. Prolonged use can lead to steroid toxicity owing to which second line drugs DMARDs such as methotrexate is an alternative, the third line of drugs being Anti TNF Inhibitors like infliximab, adalimumab. Skin lesion such as erythema nodusum resolve spontaneously in most of the cases, however during the acute phase of pain short course of NSAIDs or glucocorticoids would suffice. Joint manifestations are best resolved by NSAIDS however if they don't methotrexate or hydroxychloroquine can be prescribed [1,2,4,5]. The complications include pulmonary hypertension and end stage lung disease and mortality approximates in 5% of cases [1,6].

2. Case Report

We herein present a case of 38 years old diabetic woman who visited the Department of Medicine of Mymensingh Medical College with the complaints of fever for 1 month which was low grade without any chills or rigor, no evening rise and highest temperature wasn't recorded. She also complained of dry cough for same duration without any seasonal or diurnal variation. In addition, she mentioned of multiple joint pain involving small joints of hands and feet namely MCP, PIP and wrist joints which was not associated with any morning stiffness. She complained of generalised weakness and fatigue on exertion. On query she also mentioned of painful reddish lesion in both the shin which was gradually increasing for a few days but ultimately resolved after she took some medications from the local chemist for her condition. She noticed loss of weight gradually and her bowel, bladder habits were normal. On examination she was mildly

anaemic and her vitals were in the normal range. Examination of the respiratory system revealed no significant abnormality and other systemic examination findings were normal. She was investigated in line for TB and possible connective tissue disease RA. Following investigations her blood reports showed Hb 11.5g/dl with normocytic normochromic anaemia and increased sedimentation. Radiological findings of x ray chest revealed bilateral hilar lymphadenopathy.

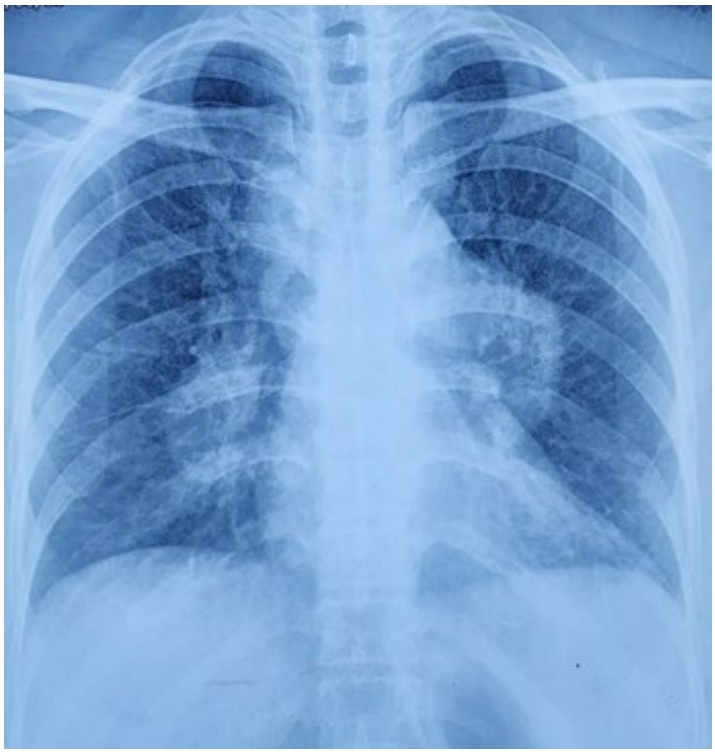

Figure 2: X Ray Chest Showing Bilateral Hilar Lymphadenopathy and HRCT Lung Revealed Bilateral Consolidation, Pulmonary Infiltrates, Bilateral Hilar and Mediastinal Lymphadenopathy

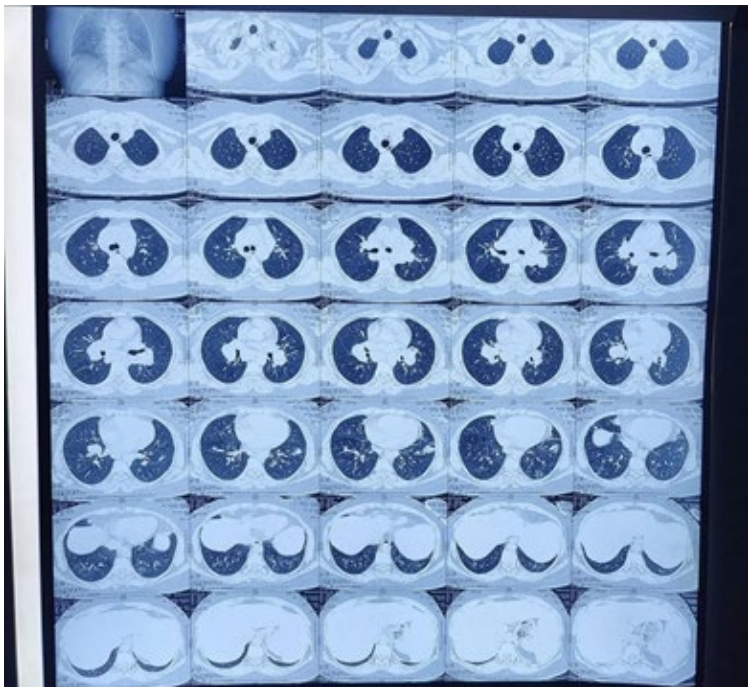

Figure 3: HRCT Lung showing Bilateral Hilar, Mediastinal Lymphadenopathy and Pulmonary Inflammatory Lesions

RA, ANA, MT serology were insignificant and sputum for AFB didn't yield any Mycobacteria (typical or atypical). ACE was higher than the normal while S. calcium and 24hrs urinary calcium were in the normal range. Spirometry revealed irreversible obstructive airway disease. She was finally diagnosed as a case of sarcoidosis and treated in line with prednisolone at the starting dose of 40mg/day which was tapered slowly over 5 months. Methotrexate was prescribed weekly and folic acid supplementation was done with it. She was advised for vaccination against H. influenzae yearly and S. pneumoniae 5 yearly. On first follow up after 3 weeks her joints pain had subsided and was doing better. She has been on regular follow up since then and is improving.

3. Discussion

Sarcoidosis is often a misdiagnosed entity in our subcontinent because of its elusive nature and prevalence of tuberculosis. As reported by Shyamal et al where Anti TB regimen was started for a patient who was suffering from Sarcoidosis; our patient complained with fever, night sweats, cough and weight loss mimicking TB. After a detailed history, clinical examination and relevant investigations he came out negative for TB, his chest X ray revealed Bilateral Hilar Lymphadenopathy and HRCT lung showed pulmonary inflammatory lesions, Bilateral hilar and mediastinal lymphadenopathy [7]. ACE level was increased which is one of the serum markers of Sarcoidosis [1]. The commonest form of arthropathy in the disease process is Lofgren's syndrome which manifests as BHL, EN and arthritis [8].

The patient had migratory polyarthritis involving small joints of hands without any morning stiffness. Articular manifestations in sarcoidosis include both acute and chronic types. Chronic form is rare which can range from non-deforming arthritis to granulomatous synovitis. Jaccoud's arthropathy can be one of the manifestations [8]. The typical clinical features of Lofgren's syndrome, increased ACE, BHL and exclusion of TB established the diagnosis. Spirometry revelaed irreversible obstructive airway disease, though sarcoidosis has been known to be a restrictive disease but as reported by Petey Laohaburanakit et al airway obstruction has become a recognised feature of disease in the recent years given it can obstruct airways at any level and when the it involves small airways it often resembles obstructive airway disease [9].

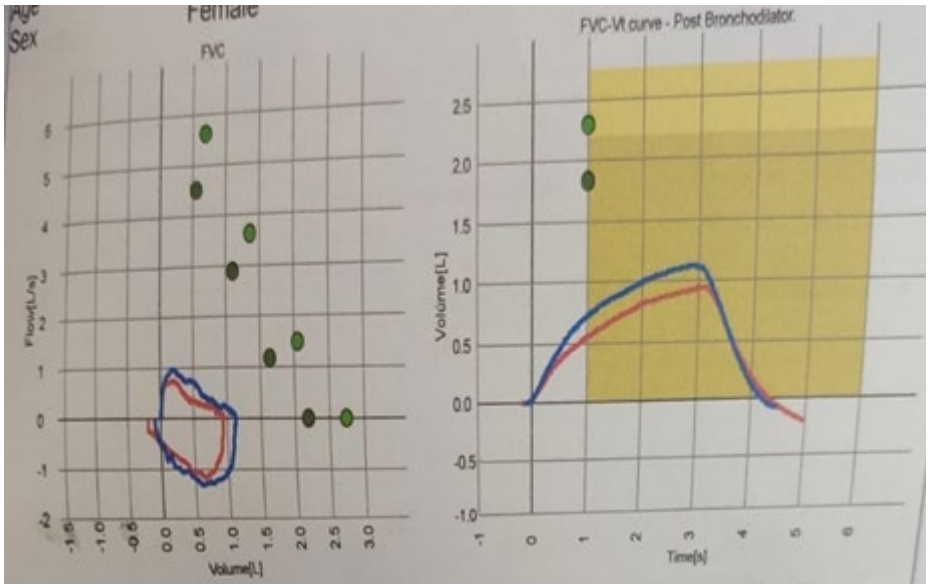

Figure 4: Spirometry showing Irreversible Obstructive Airway Disease

The treatment was started in line for Sarcoidosis with oral prednisolone 40mg/day and it was gradually tapered over the period of 5 months. Methotrexate 15mg per week with folic acid supplementation was prescribed [1,2,4,5]. She was on insulin, was asked to continue taking that and lifestyle modification for good diabetic control was advised. She was also advised to vaccinate herself for H. influenzae yearly and S. pneumoniae every 5 years. Raising awareness about this disease though rare helps the physician make an early diagnosis leading to timely treatment of the patient and improvement of the quality of life.

4. Conclusion

The main objective of the authors is to highlight a case of sarcoidosis which is a rare, elusive, often misdiagnosed disease entity in our subcontinent owing to its close resemblance to other granulomatous disease. It is crucial to suspect sarcoidosis as early treatment can be life saving for the patient otherwise delayed diagnosis can lead to life endangering complications.

Consent

An informed written consent was obtained from the patient based on journal's policy.

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