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Atypical Presentation of SLE: A Case Report

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ABSTRACT:
Systemic lupus erythematosus (SLE) is an immune complex disease with a plethora of clinical manifestations. Lymphadenopathy is a non-specific and benign manifestation of SLE, mainly seen in young patients with constitutional symptoms and reactive lymphadenitis is the most frequent finding on microscopic examination. Detailed history taking, clinical examination and laboratory investigations are needed to arrive at a diagnosis. Here we present a case of 26 year old female presenting with lymphadenopathy, fever, malaise and generalized weakness with history of multiple joint pain, initially started on Anti-tubercular therapy (ATT) empirically outside hospital and finally diagnosed as SLE.
Keywords: SLE, Lymphadenopathy, Tuberculosis, Anti-tubercular therapy

I. INTRODUCTION:

Lymphadenopathy is defined as enlargement of one or several lymph nodes[1].

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune inflammatory disease, presenting with a variety of clinical features. The diagnosis is made by the presence of four or more of the 11 criteria laid by American College of Rheumatology, none of which includefever or lymphadenopathy.[2]

We present a case of 26 year old female presenting with lymphadenopathy, malaise and fever, who was initially started on Anti-tubercular therapy (ATT) outside hospital. In view of persisting symptoms, she reported at our hospital, extensive work-up was done and final diagnosis of SLE was made.

II. CASE HISTORY:

A 26year old unmarried female presented with C/O generalized weakness and malaise since last 3 months, multiple nodular swellings around neck since last 2 months and fever on and off since last 2 month. She gave history of swelling & pain of small joints of hands 18 months back, which improved with treatment (detail not available).She gave no history of exposure to TB. She was started on ATT outside on suspicion of tuberculosis. Patient presented to our hospital for second opinion.

On examination, the patient was malnourished, pale, small joints of hand and elbow were tender with no deformity. Oral ulcers were seen on the hard palate, which were non-healing, painless and persisting since a month, as described by the patient on enquiring. There were multiple, painless, enlarged, soft, non-tender, freely movable lymph nodes in the cervical and axillary areas, the largest of which measured 3*2 cm (Fig 1).

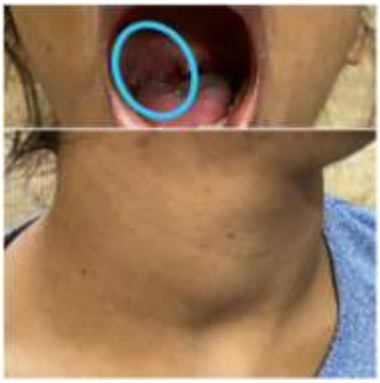

FIG-1

On investigation, complete blood count revealed pancytopenia, with Hb 6.7 g%, RBC $2.33*10^6/\mu L$, WBC $2.28*10^3/\mu L$, Platelet Count $72*10^3/\mu L$. ESR 90mm/h, Direct Coomb's Test was positive. Mantoux test was negative. LFT/KFT were normal. Ultrasound abdomen showed enlarged liver with normal echotexture, spleen & kidneys were normal.

CECT chest revealed multiple small ground glass attenuation centrilobular nodules in both lungs, more on right side. Multiple mediastinal and hilar lymph nodes were seen, some of them were calcified. Bilateral cervical and axillary lymphadenopathy was also noted, with no evidence of necrosis.

FNAC of cervical lymph node (fig 2) was suggestive of hypercellular smear with polymorphous population of lymphocytes, centrocytes, centroblasts, plasma cells,

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immunoblasts with occasional multinucleated giant cell. Few lympho-histiocytic tangles were observed, with no evidence of necrosis, Acid Fast Bacillus was also negative.

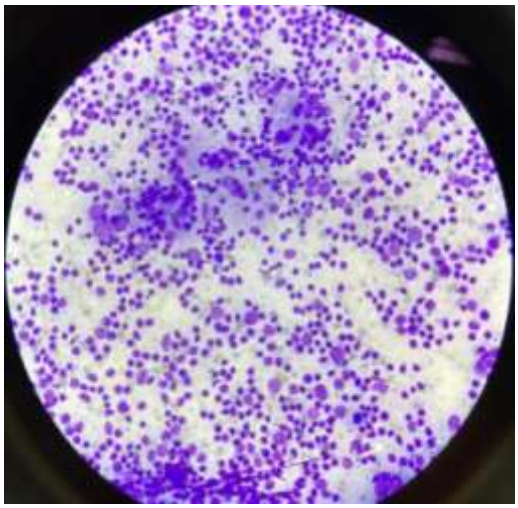

FIG-2

On suspicion of SLE, the patient was further investigated. ANA Hep-2 Positive (3+),ANTI-ds DNA Antibody was found to be 118.46 IU/ml (normal<30.00), Serum anti-Smith antibody was 123.91 units (normal <20 units), Serum SSA/Ro Ab was 168.27 units (normal <20 units) and Serum SSB/La Ab was 196.62 units (normal <20 units), Complement-3 value was 20.8 mg/dl (90-180), 24 hrs urinary protein was 0.16 gm.

The diagnosis of SLE was thus established. Anti-tubercular therapy was stopped and initially inj Methylprednisolone (500 mg) IV given for 3 days and subsequently switched to oral Prednisone (60 mg) daily, along with tab Hydroxychloroquine (200mg/d). She was also transfused with one unit of packed red cells. Patient improved symptomatically with healing of oral ulcers and regression in size of lymph nodes and doses of prednisone were decreased.

III. DISCUSSION:

Lymphadenopathy is a common clinical manifestation in outpatient departments and could represent a vast array of infectious and non-infectious etiology. In India and some other developing countries, tuberculosis (TB) is often the first differential diagnosis for a patient who presents with lymphadenopathy. However, many studies have reported that more than 50% of cases of lymphadenopathy are due to non-TB causes. Excision biopsy (EB) or Fine needle aspiration with histopathology and/or microbiological examination, is the only way to exclude TB.[3] Generalized lymphadenopathy, defined as involvement of three or more noncontiguous lymph node areas, has been found to be commonly associated with non-malignant disorders such as infections, SLE or other inflammatory diseases.[4]

SLE is a chronic multisystem autoimmune inflammatory disease with a variety of clinical manifestations, affecting mostly young females. However, generalized lymphadenopathy is not considered a criterion for diagnosis of SLE. The exact prevalence of generalized lymphadenopathy in patients with SLE is unknown.[2] In one retrospective study of 90 SLE patients, 23 patients (26%) were found to have lymphadenopathy.[5]

Although lymphadenopathy is a non-specific sign of SLE and does not co-relate well with disease activity, it may often be the first clinical manifestation. Increased constitutional symptoms have been associated with lymphadenopathy such as fatigue, fever and weight loss, more cutaneous and mucosal signs (malar rash, vasculitis, skin ulcers, mouth ulcers, discoid lesions, alopecia and subcutaneous lupus erythematosus), higher rate of hepatomegaly and splenomegaly, increased anti-dsDNA antibodies titers and decreased complement levels. The British Isles Lupus Assessment Group index has also found higher disease activity among patients with lymphadenopathy.[6]

Lupus Lymphadenopathy commonly involves cervical and axillary regions, and the afftected lymph nodes are soft, mobile, painful and not fixed to deep tissues.[7]

The histological findings of lymph node are usually nonspecific and may consist of moderate follicular hyperplasia associated with increased vascularity and scattered immunoblasts and plasma cells[8] (lymphoplasmacytic proliferation), several degrees of coagulative necrosis with hematoxylin bodies or reactive follicular hyperplasia.[9]

Apart from tuberculosis, the other differential diagnosis of lymphadenopathy syndrome in SLE includes Kikuchi-Fujimoto disease, or histiocytic necrotizing lymphadenitis, manifested by cervical lymph node involvement, fever, night sweats and leukopenia.

Several studies have reported increased risk of haematological malignancies in SLE since 1970s. Hence, the differential diagnosis of Hodgkin's or non-Hodgkin's lymphomas, acute or chronic lymphocytic leukemia, hairy cell leukemia, malignant histiocytosis and metastasis should also be kept in mind.[6]

Castleman's disease, syphilis, sarcoidosis, infectious mononucleosis (Epstein–Barr virus, cytomegalovirus), herpes simplex, human immunodeficiency virus (HIV), hepatitis B virus (HBV), hepatitis C virus (HCV) are among the other differentials that may also present with lymphadenopathy[10].

IV.CONCLUSION:

This is a rare case of SLE with atypical presentation. Another interesting aspect is that there was no history of photosensitivity, discoid lesions, malar rash etc. as commonly seen in typical cases of SLE. A careful medical history, proper examination and investigations may point towards the definitive diagnosis. As in our case, fever and lymphadenopathy raised a suspicion of TB, however, the patient being young in her reproductive years with no history of exposure to TB, presence of oral ulcers and involvement of joints challenged the initial diagnosis and after detailed investigations, diagnosis of SLE could be established

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