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"SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING WITH SCHUSTER SIGN: 2 CASE REPORTS FROM A TERTIARY CARE CENTRE IN UPPER ASSAM"

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ABSTRACT Systemic Lupus erythematosus is a multi system auto immune disorder involving auto antibodies with varying presentations ranging from musculoskeletal manifestations to life-threatening Multi organ dysfunction. Some Patients initially present to primary care physicians with symptoms such as Hyperpigmented rash over ears , which is often misdiagnosed and resulting in delayed referral to Rheumatologist and delayed treatment which can have negative impact on patient quality of life and disease burden. Systemic lupus erythematosus may not always present with typical malar rash and joint pain as seen commonly , Hyperpigmented lesions with central scarring over conchal bowl of the ears described in literature as 'Schuster sign' is an important dermatological manifestation of Discoid Lupus, which is present in 30% cases of DLE . It is Scarring due to chronic inflammation, Which if not treated early can progress to Systemic lupus erythematosus in about 5% to 25 % in a duration of months to few years[1]. We briefly discussed two cases with 'Schuster sign' who attended our Rheumatology OPD and later underwent detailed evaluation and diagnosed to have Systemic lupus erythematosus. This involves two young females who had primary complaints of Hyperpigmented Rash over ears, which was missed in Early part of patient care. SLE was suspected when we took detailed history and Clinical examination. Thorough knowledge regarding Dermatological Manifestations of Lupus is essential for early Diagnosis and treatment. Hence This case provides insight into a very important sign of 'Schuster sign' which if picked up early will reduce the morbidity and mortality associated with the disease[1]. A strong clinical suspicion of Lupus should be kept in mind while encountering patients with such findings[2].

KEYWORDS : Systemic Lupus Erythematosus , Schuster sign , DLE , Discoid Lupus Erythematosus

INTRODUCTION

Systemic lupus erythematosus is a Multi system autoimmune disorder involving innate and adaptive immune system activation , T cell and B cell activation , increased expression of interferon gamma and release of auto antibodies which results in antigen antibody complex formation and deposition in joints , blood vessels and kidney resulting in varying manifestations such as arthritis, vasculitis and nephritis[3]

The clinical phenotype is highly heterogeneous, ranging from mild cutaneous and musculoskeletal involvement to life-threatening renal, neuropsychiatric, or cardiopulmonary manifestations. The disease course is variable with relapse and remission episodes[4]. Patient may not always present with typical symptoms of SLE such as joint pain or alopecia or malar rash .Understanding the diverse manifestations of SLE is essential for timely recognition, diagnosis, and management across Rheumatology and internal medicine practice[5]. 'Schuster sign' , though classically associated with other clinical contexts, can reflect systemic inflammatory burden or localized pathophysiology relevant to autoimmune disease activity and warrants careful documentation when observed in SLE[6]. This case highlights the importance of thorough clinical examination and comprehensive evaluation to diagnose and treat early , inorder to reduce the complications associated with.

CASE PRESENTATION 1

A 15 year old Young girl hailing from a small village, studying in 9th standard from Upper Assam , India presented to our Out Patient Department, Rheumatology, Assam Medical College Dibrugarh with Primary complaints of fatigue and discolouration of skin over bilateral ears for past 12 months duration , she also had low grade fever for 3 months which persisted throughout the day not associated with chills and rigor . She had joint pain associated with swelling involving bilateral wrist , elbow , knee , metacarpo phalangeal , proximal and distal interphalangeal joints . Patient slowly developed bilateral pedal edema and Abdominal distension over 1 month . There was no history of cough or TB contact . No history of altered sensorium . No history of rash over cheeks.

On examination BP 100/60 mm Hg Pulse Rate 80 per minute Spo2 99% under room air . Chest right sided basal air entry decreased . Per abdomen distended , shifting dullness present. GCS E4V5M6 , S1S2 present , No Murmur .

On Evaluation , Hb 9.0 total leucocyte count 5370/mm³ platelet count 1.8 Lakhs, creatinine 1.2 , ESR 40(elevated) CRP less than 0.5 . Low C3 C4 levels , pANCA cANCA negative, Hepatitis B Hepatitis C HIV 1 & 2 were negative, Mantoux test Negative, Albumin globulin ratio reversed , serum albumin 2.6 gram/dL . Serum Triglycerides was raised 351 mg/dL . Urine analysis showing protein 2+ . 24 hour urine protein 2.6 gram per 24 hours . ANA PROFILE report showing 3+ intensity, 1/360 Titre, Antibodies against dsDNA , Smith, Nucleosome, histone positive . Chest xray PA view showing right sided minimal pleural effusion . USG guided Pleural fluid analysis showing 20 cells lymphocyte predominant, protein, ADA levels and sugar levels normal.

Diagnosis was made as Systemic lupus erythematosus with Lupus Nephritis.

Patient was managed with Methyl prednisolone pulse dose 15mg/kg/day for 3 days followed by oral prednisolone 1mg/kg/day followed by tapering dose . She was also intiated on Tab Hydroxychloroquine 200mg at bed time , Tablet Losartan 50 mg once daily, Tablet cotrimoxazole 960mg alternate day , proton pump inhibitors , Calcium and vitamin D3 tablets , Sun screen lotion and Diuresis with 20% Human Albumin.

She was started on Tablet MMF 2 gram per day and discharged at day 6 of admission . After two month follow up, patient was doing well . Edema reduced significantly , Rash over ears resolved after 2 months gradually.

This case gives importance of knowing the skin manifestations in Lupus to diagnose and treat the cases early in the course of disease .

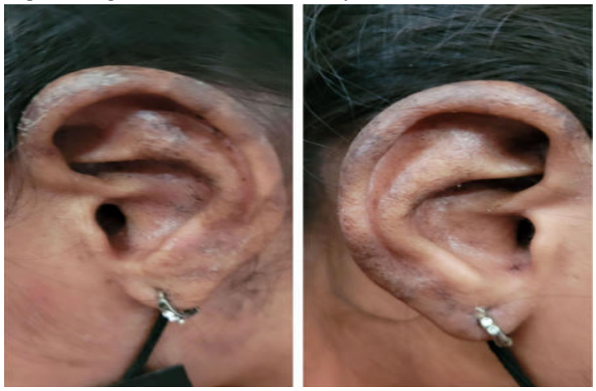

Figure 1: Bilateral Ear Showing 'Schuster Sign'

CASE PRESENTATION 2

18 year old Female , Studying in Higher secondary school in Assam , presented with complaints of fever for 3 weeks , fatigue , joint pain and Hyperpigmented Rash over bilateral ears and erythematous rash over Malar area, mild bilateral pedal edema for 7 days.

On taking detailed history, malar rash occurred along with the fever of 3 weeks duration . But Hyperpigmented lesion over bilateral ears were present for past 2 years , which they neglected as patient was Asymptomatic in those 3 years . No history of abnormal jerky movements, tongue bite , involuntary micturition or defecation, altered sensorium.

As patient presented with 'Schuster Sign' and prolonged fever with bilateral symmetric polyarthritis , we evaluated the patient as a suspected case of systemic lupus erythematosus.

On Evaluation , Hb 5.6 , ferritin 656 , ESR 120 , CRP 0.5 , total leukocyte count 6500 , platelet count 1.5 Lakhs , creatinine 0.9 , Serum Albumin 4.0 , Serum Globulin 3.1, urine RE albumin NIL , 24 hour urine protein 525 mg / 24 hours , C3 C4 low , pANCA , cANCA Negative . Hep B Hep C , HIV Negative , Mantoux test Negative .

ANA Profile Showing Antibodies positive against RNP 3+ , Smith 3+ , Histone 2+, Ribosomal protein 2+ , Nucleosome 2+ .

Diagnosis of SLE with Lupus Nephritis with Severe anemia of chronic disease was made .

Patient was initiated on Inj Methylprednisolone 750mg per day followed by oral prednisolone 20mg per day , tapered in subsequent visits. She was also started on Tablet Hydroxychloroquine 200mg at bed time , Tablet MMF 500mg two tablets twice daily , Tab losartan 25mg one tab twice daily , calcium tablet once daily , proton pump Inhibitors , Tab Cotrimoxazole 960mg one tablet alternate day , sun screen lotion to apply over sun exposed areas .

After 3 month of follow up , Malar Rash resolved completely. However , Hyperpigmented lesion resolved partially, but not fully . Patient's fever subsided , joint pain resolved , general condition improved .

DISCUSSION

Schuster sign' is an importat sign of Lupus , which is to be kept in mind while dealing with patients of prolonged fever , joint pain , Alopecia . We discussed 2 cases presenting with 'Schuster Sign' which is often seen in 30% cases of discoid lupus erythematosus , with progression to Systemic Lupus erythematosus in 5 to 25% of cases[1].

So , Thorough knowledge of Dermatological Manifestations in Lupus is essential for early diagnosis and subsequent treatment. It helps in reducing the morbidity and complications associated with the disease and improved outcome.

CONCLUSION

Schuster sign is an important sign in SLE , clinical features and thorough clinical examination can aid in early Diagnosis and management thereby reducing the mortality and morbidity associated with the disease. Thereby preventing or slowing the progression of disease and Life threatening Multi organ involvement.

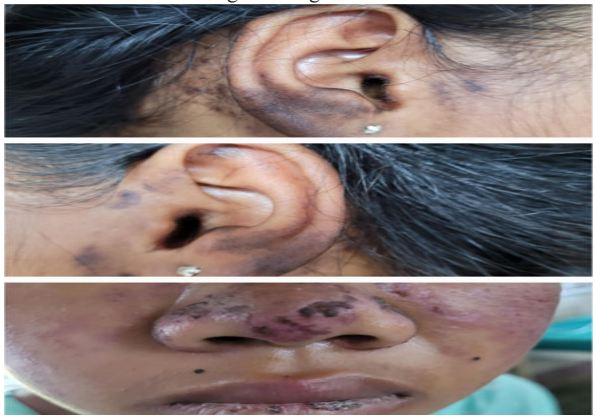

Figure 2 : Hyperpigmented Lesion Involving Bilateral Ears And Malar Rash

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