Can Mumps present as Lupus storm?

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Abstract:

Systemic lupus erythematosus is an autoimmune multisystem disease ,common in female gender. There are diversities in presenting manifestation of SLE like cutaneous, musculoskeletal, constitutional, neurological, renal, lymphadenopathy, vasculitis. Derangement of the normal processes like sequestration of self antigens, tolerance of relevant T or B cells predispose to the development of autoimmunity. These abnormal responses relate to stimulation of exogenous agent like viral agents after which autoimmune disease manifest. We report a rare case SLE clinically manifested after acute viral parotitis or mumps. This 18 year girl had never experienced lupus activity in the past and clinically manifested as acute flare of lupus only during the convalescent phase of mumps .

Introduction

SLE is an multiorgan autoimmune disease which is characterized by presence of autoantibodies and immune complexes.SLE manifest atypically in up to one-third of cases.The atypical manifestations of SLE can be parotitis, pericarditis, pancreatitis or hepatosplomegaly.[1]Parotitis per se occurs in 2.5 % cases of SLE presenting atypically.[2] We discuss whether the parotitis in our case was a separate entity or part of autoimmune disease.We also discuss the association between acute viral infections and SLE, as the acute viral infections have long been suggested to induce autoimmune responses although the mechanism by which viruses cause generation of autoantibodies is still obscure

Case History

18 year old girl presented to us with history of high grade fever, bilateral swelling over parotid region, ear ache since 8 days. She also had difficulty in swallowing. She had no previous history of mumps, and vaccination status for mumps in the childhood was not known. On clinical examination, she had fever, pallor, tachycardia, bilateral symmetrical parotid swelling obliterating the space between the ear lobe and the angle of mandible. Both the parotid swellings were tender. Rest of systemic examination was normal. With these findings there was no doubt in clinical diagnosis of mumps. The patient was started on symptomatic therapy. The complete blood count, blood urea and serum creatitine were within normal limit. Her ELISA for HIV and serum HbsAg were negative. Her parotid swelling gradually decreased. Also the range of fever was reduced.

However after 10 days of admission when the parotid swelling started subsiding, she acutely developed butterfly rash over face and multiple macular eruptions over trunk and extremities. Her rash was extensive and was so severe that she developed multiple ulcers over face, shoulder and hands .Also she developed oral ulcers. Her concurrent serological investigations showed positive anti nuclear antibody(ANA) test and positive anti double stranded DNA(dsDNA) with titer 135 IU/ml.ESR was 45 mm/hour, spot urine examination was positive for albumin and cellular casts on microscopy,24 hours proteinuria 1.2 g/day,blood urea 61 mg/dl, creatinine 1.6 mg/dl . Her repeat complete blood counts were normal , Hb was 9.5gm% .Her X ray chest PA view did not reveal any abnormality. She had persistent moderate grade fever and persistent tachycardia . Her ECG was showing sinus tachycardia and 2D echocardiography was normal . Her thyroid hormonal estimation was normal .So on the basis of these findings as per the American college of Rheumatology ,the diagnosis of SLE was established .
The patient was started on IV Methylprednisolone 1 gm

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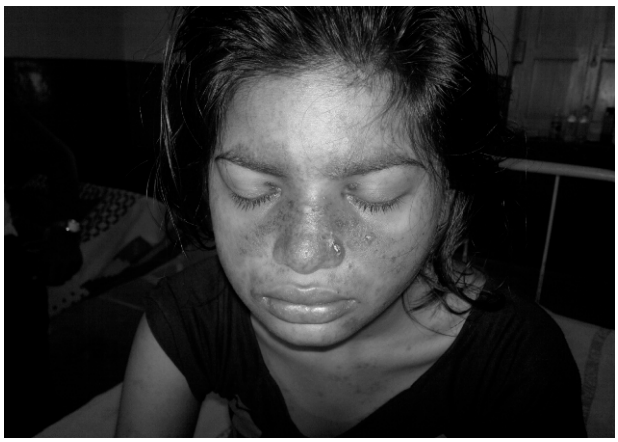
daily for 3 days followed by oral prednisone 40 mg OD,oral Hydroxychloroquine 200 mg BD, Tab Azathioprine 75 mg OD, topical antibiotic cream for local application, sunscreen lotion for local application.With this treatment for about 4 weeks, her rash faded, ulcerated lesions healed, fever completely subsided,24 hours proteinuria decreased to 0.3 g/day,her blood urea and serum creatinine became normal. After 4 weeks of treatment ,dose of Hydroxychloroquine was reduced to 200 mg OD, tab Prednisone tapered to 30 mg OD, tab Azathioprine continued with 75 mg OD. With this treatment ,patient was discharged and advised for follow up. Serum mumps IgM was done during discharge and was negative i.e 5.85 IU ,positive value being >11 IU.

As per WHO criteria for lupus nephritis ,it is mandatory to have renal biopsy for diagnosis of lupus nephritis,but because we could not obtain consent of patient and her parents despite repeated explanation,renal biopsy was not done.Patient is still under regular follow up.

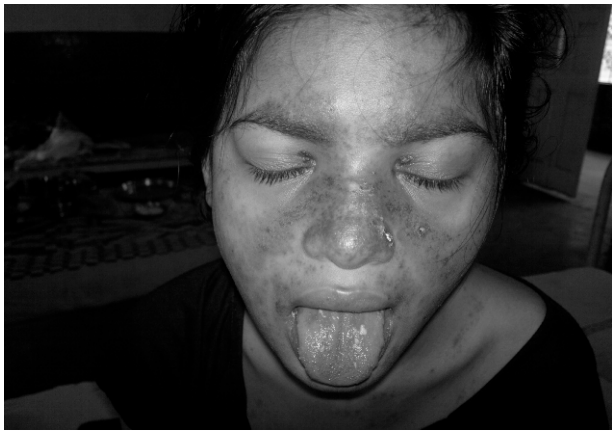
Discussion

There are various causes of parotitis like infective and non infectious. The common infective causes of parotitis are mumps caused by paramyxovirus, others being coxsackievirus, parainfluenza virus,HIV virus. The systemic noninfectious causes of parotitis are Sjogren syndrome,sarcoidosis,diabetes mellitus,uremia.SLE is rarely associated with parotid gland enlargement. The atypical manifestations of SLE can be parotitis, pericarditis, pancreatitis or hepatosplomegaly.[1] Parotitis per se occurs in 2.5 % cases of SLE presenting atypically.[2] In such cases ,the parotitis was mostly unilateral, of longer duration and accompanied with lupus activity in other organs.[3] In such cases,parotitis was resolved with drug induced remission of lupus. In some cases, parotitis associated with lupus activity was acute in onset, either unilateral or bilateral , and resolved shortly after treatment for lupus.[4] The FNAC of the gland were showing lymphoplasmacytic infiltrate in those cases.[5] In our case, the patient initially presented with acute bilateral parotitis, clinically diagnosed as mumps without concomitant lupus activity. The parotitis subsided completely within short period of 10 days followed by sudden flare up of lupus.So in our case it is debated whether the parotitis was due to infective cause or part of autoimmune disease. We strongly believe that mumps was the cause of acute bilateral parotitis in this case and was not per se manifestation of SLE though the serological diagnosis of Mumps could not be established. It may probably because serum IgM for mumps was done 4 weeks after parotitis, and the sensitivity of test in fourth week is very less and negative IgM does not rule out mumps as per CDC guidelines.[6] FNAC of the parotid gland was not done as there was no doubt of clinical diagnosis of mumps. We believe that mumps may have induced the lupus disease clinically after subsiding the parotid gland swelling. Epstein-Barr virus and the Coxsackie virus were the previously known triggering factor for active lupus activity. In this case mumps may have acted as triggering factor for flare up of lupus disease.

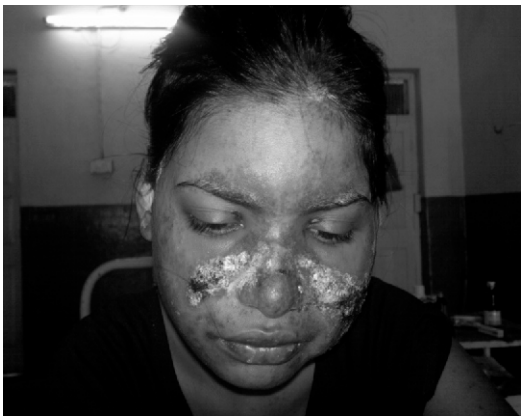
1.Day 1O of admission



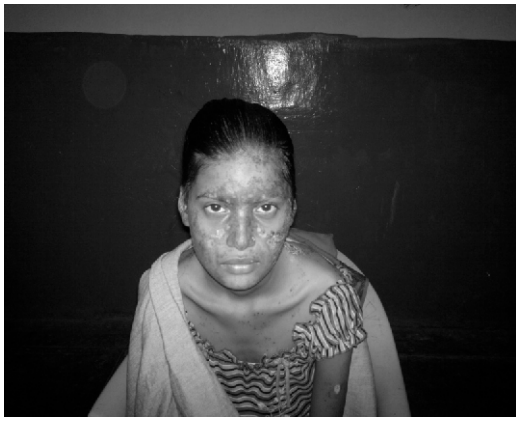
2.Day 11 of admission



3. Evolution of rash over next 7-8 days



4.Day of discharge



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