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13 YEAR OLD PATIENT WITH LUPUS RESISTANT TO MAXIMUM IMMUNOSUPPRESSION.

CARDIOLOGY & RHEUMATOLOGY

KEYWORDS: SLE, Lupus nephritis, immunosuppression

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ABSTRACT

Juvenile SLE is an autoimmune disease with multisystem involvement leading to inflammatory damage to joints, heart, lungs, kidneys and central nervous system. We are presenting the case of a patient with recurrent admissions to the general medicine ward for fever who was eventually diagnosed as having juvenile systemic lupus erythematosus (SLE). Despite maximum immunosuppression, patient succumbed due to concomitant neurological, cardiovascular and renal system involvement.

Content: A 13 year old girl, the first born child of a non consanguinous marriage was admitted to the general medicine ward for the fifth time during previous six months. The first four admissions were for fever. Each time, patient would present with a history of mild to moderate grade, intermittent fever not associated with headache, vomiting, cough, sore throat, abdominal pain, burning micturition or rash over body. Birth, growth and developmental history were unremarkable. Each of the four times, the patient was treated with fluids and anti-pyretics and discharged with a diagnosis of viral fever owing to the negative work up for pyrexia in each admission. The fifth time, patient presented with complaint of dyspnea since four days. It was not associated with fever, cough or chest pain. Physical examination revealed pallor, a rapid feeble pulse and hypotension associated with bilateral pedal edema. Complete blood count showed anemia and lymphopenia. Renal and liver function tests were normal. ECG showed sinus tachycardia. Chest X Ray showed gross bilateral pleural effusion. Thoracocentesis revealed transudative pleural effusion. Ultrasonography of the abdomen showed kidneys with normal size and echogenicity and gross ascites. 2D Echo showed severe left ventricular dysfunction with an ejection fraction of 20%.The valve morphology was normal. Cardiac enzymes (creatinine kinase and cardiac troponins) were elevated. Urine routine and microscopy showed heavy (3+) proteinuria with granular casts. 24 hour urinary protein measured 2400 milligrams. ANA (Anti nuclear antibody) was +3 positive, nuclear homogenous pattern. ANA blot was sent which came back as strongly positive for SLE. The patient was diagnosed as juvenile lupus with myocarditis with plan for renal biopsy. Patient was started on full dose immunosuppression in the form of injectable methylprednisolone and GDMT for congestive cardiac failure. One week into the treatment, patient had single episode of generalized tonic clonic seizure which lasted for ten seconds. It was followed by 4 episodes of seizures during the time she was admitted in the hospital. Neuroimaging was suggestive of demyelination. CNS Lupus was added to her diagnosis. She was managed for the same with immunosuppression and broad spectrum antibiotics to cover secondary infection. After she recovered from the acute phase of the CNS lupus, renal biopsy was done which showed class 4 lupus nephritis with diffuse subendothelial immune deposits and mesangial alterations. dsDNA was positive and serum C3 levels were very low. Patient was started on cyclophosphamide pulse therapy according to her body weight every 15 days for a total of 6 doses along with continuation of anti failure treatment. Her 6 month follow up cardiac function improved to an ejection fraction of 45%. However she kept having minor flares. She was given a trial of azathioprine as well as mycophenolate mofetil. Patient was also administered monoclonal antibody in the form of 500 milligrams of Rituximab but her flares never completely resolved. Fifteen days later patient was brought dead in the emergency room.

Discussion- There is no cure for SLE but proper and timely treatment can help the patient significantly. Nearly 20% of the patients of SLE have disease onset before the age of 20 years but it is rare before the age of 5 years. Compared to adult onset SLE, juvenile SLE which is defined as disease onset less than 18 years of age, is more severe with more organ involvement and higher mortality. Like the adult onset SLE, majority of the patients in the juvenile group are female. Renal, cardiac and central nervous system involvement cause significant morbidity and mortality. Patients with juvenile onset SLE more often have renal involvement and encephalopathy than patients with adult onset SLE. Anti-ribosomal P, Anti-ds DNA and antihistone antibodies are more often found in patients with juvenile SLE.

Cardiovascular disease is a frequent complication of SLE and may involve pericardium, myocardium, valves and coronary arteries. Myocarditis is uncommon in SLE which our patient had.

Our patient had very severe disease with very rapid involvement of all the major systems namely renal, cardiovascular and central nervous system within a span of less than one and a half years. She showed no remission despite maximum immunosuppression and most likely succumbed to fatal arrhythmia.

References :

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