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A YOUNG LADY WITH REFRACTORY LUPUS: TRIALS AND TRIBULATIONS

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INTRODUCTION

Systemic lupus erythematosus (SLE) is a multisystem, auto- immune disorder, characterized by the presence of multiple autoantibodies and deposition of immune complexes in various tissues **[1]**.

Presentation

22-year-old lady known case of Hypothyroidism, presented in 2018 September with fever polyarthritis seizures and small vessel vasculitis on leg. She was evaluated and diagnosed as Systemic lupus erythematosus with hematological and neurological and vascular involvement

Initial evaluation revealed pancytopenia with lymphopenia very low complements and positive ANA, dsDNA, Sm and ribosomal antibodies.Her direct coombs test was also positive. She was managed with mycophenolate mofetil and pulse steroids followed by tapering dose of steroids starting from 1mg/kg and hydroxy-chloroquine. She was maintaining clinical and laboratory based remission for 6 months.

Course and Management

However, she developed chronic diarrhea and cryptosporidiosis with MMF and hence azathioprine was substituted. She developed

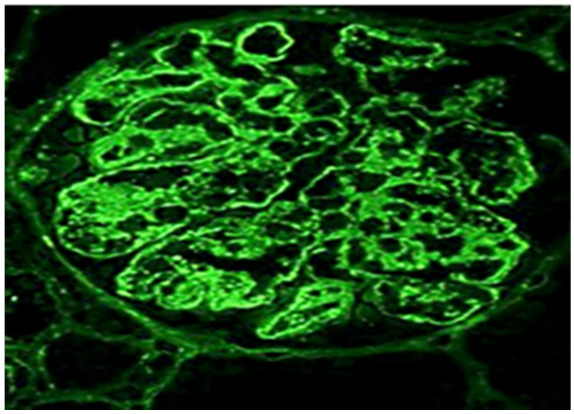
severe pancytopenia due to azathioprine and evaluation revealed TPMT positive. Hence azathioprine was discontinued. Meanwhile, she used to have persistent low mcv anemia with decreased RDW and normal iron studies. Hb electrophoresis was suggestive of hemoglobinopathy.

In January 2020, she developed anasarca with fluid overload following a viral infection.Evaluation revealed polyserositis with azotemia and nephrotic range proteinuria. Her APLA was positive. Renal biopsy was done revealed class 4 lupus nephritis with interstitial nephritis. She was given pulse methylprednisolone followed by IVIG and multiple doses of parenteral albumin in view of proteinuria. As she was unmarried parents and the patients refused cyclophosphamide and also embryo preservation procedure. During the course of illness,she noted to have severe vitamin D deficiency and dysglycemia with prolonged steroid therapy.

IMAGES

In view of the above she was started on Rituximab for refractory lupus, however after 2 doses she developed multiple folliculitis. Evaluation revealed neweronset severe hypogammaglobulinemia, which persisted for 3 months. Further dose was declined due to COVID pandemic. Presently she is on tacrolimus with MMF underclose supervision though symptomatically better she continues to have lymphopenia with high dsDNA levels Nephrotic range proteinuria with severe Hypoalbuminemia

Final diagnosis of refractory lupus with class 4 lupus nephritis/tpmt deficiency/ crytosporidiosis treated/rituximab induced hypogamma globulinemia was made.



DISCUSSION

Nephritis, which is the most frequent serious manifestation of the SLE can affect up to 60% of adults. Introduction of corticosteroids and then immunosuppressive therapies, have improved prognosis, such that 5-year survival rates are approximately 95%, and at 10 years 90%. Rates of end-stage renal failure (ESRF) have, however, remained static at 10–20% despite the effective therapies available. Various biologics have been tried in patients with relapsed or refractory lupus nephritis **[1]**.

CONCLUSION

Refractory nephritis develops in a small subset of patients with SLE. Multiple therapeutic options are currently available, the trials and tribulations of both patients and consultants are highlighted here which can be overcome with specialized dedicated and high-quality rheumatic care.

REFERENCE

[1] Smith RM, Clatworthy MR, Jayne DR. Biological therapy for lupus nephritis—tribulations and trials. Nat Rev Rheumatol., 2010; 6(9): 547.