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LUPUS PROTEIN-LOSING ENTEROPATHY (LUPLE): DESCRIPTION OF THIS RARE MANIFESTATION IN SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) AND A REVIEW OF LITERATURE

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BACKGROUND

Protein losing enteropathy (PLE) is characterized by the leakage of serum proteins from the gastrointestinal tract leading to a generalized edema and hypoalbuminemia without proteinuria. Lupus protein-losing enteropathy (LUPLE) is a rare manifestation in SLE and its diagnosis requests exclusion of renal manifestations.

CASE REPORT

Female 25 years old patient with a prior diagnosis of SLE since 2015 due to anasarca, malar rash, alopecia, arthralgia, decrease in complement levels, positive FAN nuclear speckled pattern 1/1280, when prednisone was started. In 2017 she was admitted in our hospital due to severe pain, paresthesia, decreased sensitivity and temperature in lower left limb, associated with intermittent claudication since the day before. This condition was also associated with nocturnal paroxysmal dyspnea without fever and other infectious symptoms. She was taking prednisone 5mg/day, Enalapril, and Simvastatin. An acute arterial obstruction diagnosis was confirmed and fasciotomy and thromboembolectomy were perfomed. Computed tomography angiography of the lungs demonstrated a pulmonary thromboembolism. At admission, she had a severe anasarca and hypoalbuminemia (0.9 g/dL), associated to decrease of lymphocytes and complement levels. Renal function was normal and no evidence of proteinuria/nephrotic syndrome was detected. Moreover, intestinal vascular and disabsorptive causes as well as hepatic dysfunction were excluded. A low Alpha 1 antitrypsin fecal and an intermediate serum levels supported the hypothesis of a protein-losing enteropathy due to lupus activity. During hospitalization, she received a methylprednisolone pulse of 1 g/day for 3 days, associated with venous albumin and loop diuretic for 10 days. Intravenous methylprednisolone was sustained in association with partial parenteral nutrition for 20 days due to malnutrition and severe hypoalbuminemia. Besides this treatment, she also presented a posterior tibial occlusion due to severe vasculitis of some distal phalanges and an amputation of one of them was performed. Immunosuppression with IV Cyclophosphamide 500mg was associated to prednisone 60 mg/day and enoxaparin 40 mg 12/12h. After this period, serum albumin levels return to normal range with complete resolution of anasarca, laboratorial parameters and with stable renal function.

CONCLUSION

LUPLE is a rare manifestation of SLE that can occurs as an initial manifestation but also during treatment, in association with other severe clinical conditions. Since it can lead to organ damage, aggressive immunossupressive treatment is necessary to get a high

response rate.