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SYSTEMIC LUPUS ERYTHEMATOSUS OVERLAID ACUTE INTERMITTENT PORPHYRIA

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BACKGROUND

Acute intermittent porphyria is a rare metabolic disorder resulting from a partial deficiency of the heme biosynthetic enzyme porphobilinogen deaminase. Development of symptoms is affected by a variety of exacerbating factors. The association between systemic lupus erythematosus and porphyria are unknown, some authors proposed that porphyria can trigger an immune response favouring SLE. Excess of porphyrins cause activation of the complement system and increases neutrophil chemotaxis when exposed to ultraviolet rays. Porphyrins stimulate antigens that promote antibody formation.

CASE REPORT

A 32 year old woman without comorbidities was hospitalized for diffuse abdominal pain, asthenia and progressive muscle weakness, progressing to quadriplegia in 2012. Patient was initially investigated and treated for Guillain Barré syndrome but she had no response after IV immunoglobulin. SLE Was suspected porphyria after 2 samples of urinary porphyrinogen positive. Due to prolonged hospitalization, she developed gluteus and ischium bedsores with frequent hospitalizations due to local infections. Patient always had a daily fever. In May 2019 she had new hospitalization due to sacral ulcer infection, requiring antibiotic IV treatment and surgical debridement. During this period she developed bilateral wrist arthritis. Alopecia and face photosensitivity was identified. Suspected of systemic lupus erythematosus with the following positive laboratory findings: ANA speckled pattern 1: 640, complement intake (C4 9.4, with reference value 17.5 - 52.2 mg/dL), direct coombs positive. Treatment with hydroxychloroquine 400mg/ day was started from Monday to Friday with an early outpatient follow-up plan.

CONCLUSION

The association between intermittent acute porphyria and systemic lupus erythematosus is rare. This overlapping demands prudence from the physician, given the chance of massive porphyrinuria after initiating some classes of drugs like high-dose antimalarials, can potentially cause life-threatening complications.