

Chorea as the first manifestation of systemic Lupus erythematosus: a case report

Coréia como a primeira manifestação do Lúpus eritematoso sistêmico: um relatório de caso

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ABSTRACT

Systemic Lupus Erythematosus (SLE) is a disorder of the immune system with a wide range of symptoms. The involvement of kidneys, lungs and central nervous system (CNS) denotes a more severe character and worse prognosis. Several neurological and psychiatric manifestations may be present, the most common being convulsion, neuropathies and acute confusional state, but chorea is seen in less than 2% of patients. Recent studies show that antiphospholipid antibodies (aPL) correlate with the appearance of chorea in SLE.

Keywords: systemic Lupus erythematosus, Chorea, central nervous system, antiphospholipid antibodies.

RESUMO

Lúpus eritematoso sistêmico (LES) é um distúrbio do sistema imunológico com uma ampla gama de sintomas. O envolvimento de rins, pulmões e sistema nervoso central (SNC) denota um caráter mais severo e pior prognóstico. Várias manifestações neurológicas e psiquiátricas

podem estar presentes, sendo as mais comuns a convulsão, neuropatias e estado de confusão aguda, mas a coreia é vista em menos de 2% dos pacientes. Estudos recentes mostram que os anticorpos antifosfolípidos (aPL) se correlacionam com o aparecimento da coreia no LES.

Palavras-chave: Lúpus eritematoso sistêmico, Coreia, sistema nervoso central, anticorpos antifosfolípidos

1 INTRODUCTION

Systemic Lupus Erythematosus (SLE) is a disorder of the immune system with a wide range of symptoms. The initial condition usually presents with constitutional symptoms, which may be accompanied by arthralgias or arthritis. The involvement of kidneys, lungs and central nervous system (CNS) denotes a more severe character and worse prognosis. Several neurological and psychiatric manifestations may be present, the most common being convulsion, neuropathies and acute confusional state, but chorea is seen in less than 2% of patients, this being characterized by extrapyramidal movements, with spasmodic, abrupt and involuntary movements. Recent studies show that antiphospholipid antibodies (aPL) correlate with the appearance of chorea in SLE. Next, we will bring the case report of a patient whose first manifestation of SLE was chorea associated with the skin condition.

2 CASE REPORT

ZCRG, female, 22 years old, referred to the CHS for presenting choreic movements and malar rash. She reports that six months ago she started a picture of uncoordinated movements in her right upper limb and malar rash associated with photosensitivity. He looked for UBS and was prescribed clonazepam. Patient progressed progressively with worsening of movements, generalizing to upper and lower limbs, typical of chorea. On physical examination, the patient was discolored +/4+, skin: malar rash, neurological: slurred speech, uncoordinated movements, conscious and oriented. Other systems without changes. Laboratory tests on admission: CBC and renal function without changes, PCR: 54.9, 24-hour proteinuria: 600mg/24h, Negative serology for viral hepatitis, VDRL and HIV. 1:320 fine speckled nuclear standard FAN, lupus anticoagulant (LAC) positive. Diagnostic hypothesis: SLE neurological and skin activity. Haloperidol 1mg/day, hydroxychloroquine 400mg/day and prednisone 60mg/day were started. The patient evolved with an improvement in the initial clinical condition, being discharged from the hospital. Returns to the rheumatology clinic in 2 months, no longer presenting malar rash and chorea, with a reduced dose of corticosteroids.

3 DISCUSSION

The involvement of SLE in the CNS still represents a great challenge in clinical practice and its pathophysiology is not completely known. Differential diagnoses should always be considered in these cases: infectious, degenerative, metabolic disorders or medication use. Some studies show that these syndromes may be associated with the presence of antiphospholipid antibodies and that thrombotic events would be related to their pathophysiology. Chorea is a rare event and mainly affects women in their teens. The causes of these events are the result of microvasculopathy, thrombosis, presence of autoantibodies or the involvement of inflammatory mediators, and the proposed treatment is based on the use of corticosteroids and symptomatic therapy with dopamine antagonists.

4 CONCLUSION

Chorea is a rare manifestation in SLE and a condition that is difficult to diagnose. Differential diagnoses should always be considered in this case. In addition, the association with aPL seems to be well established. Early diagnosis, as well as adequate treatment, promote a good quality of life for the patient.

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