SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
FORTALEZA 2019
PROMOÇÃO
70 anos Sociedade Brasileira de Reumatologia
REALIZAÇÃO
SCR SOCIEDADE CEARENSE DE REUMATOLOGIA

PRIMARY DIFFUSE LARGE B-CELL LYMPHOMA OF THE CENTRAL NERVOUS SYSTEM MIMICKING BEHÇET'S DISEASE

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BACKGROUND

Behçet's disease (BD) is a systemic vasculitis typically characterized by painful oral and genital ulcers associated with cutaneous, ophthalmologic, gastrointestinal, neurological, vascular and joint involvement. It´s a disease that presents multiple differential diagnoses due to its systemic character and absence of specific diagnostic modalities. Next, we present a case of a patient who had the final diagnosis of Lymphoma despite multiple manifestations compatible with BD.

CASE REPORT

A 61-year-old female patient, previously hypertensive and with recurrent oral ulcers is admitted into the emergency room with complaints of low visual acuity associated with paresis and paresthesias in the upper and inferior left limbs, which started 4 months prior, with worsening one week before. Neurological examination showed mental confusion associated with spastic hypertonia in the left upper limb. Magnetic resonance imaging (MRI) of the skull revealed multifocal involvement of the cerebral white matter and the deep gray nuclei, suggesting the hypothesis of an inflammatory process with a characteristic pattern of vasculitis (Figure 1), excluding infectious and demyelinating processes. Evaluated by ophthalmology, she was diagnosed with intermediate and posterior uveitis. The findings of cerebrospinal fluid analysis were discrete increase of proteins and leukocytes; infectious screening and Patergia Test negatives. Faced with the clinical and imaging findings of the patient a BD hypothesis has been performed. It was started pulse therapy with methylprednisolone for 5 days (total 5g) with an improvement of symptoms. She was discharged on the use of prednisone 1 mg/kg/day and azathioprine 200 mg/day. She returned to the emergency room 3 weeks after discharge with a worsening of the neurological condition. A new image examination revealed an increase of the old lesions and appearance of new cortico-subcortical lesions, corpus callosum and slight involvement of the brainstem (Figure 2). A stereotactic biopsy of the central nervous system was performed and the result was compatible with primary Diffuse Large B-Cell Lymphoma of the central nervous system.

CONCLUSION

Premature diagnostic closure is one of the major factors leading to an error in clinical practice. Before considering the initiation of BD´s treatment, the rheumatologist should ensure that all differential diagnoses have been considered and ruled out.