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PEDIATRIC BEHÇET DISEASE AND BRAIN INVOLVEMENT

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BACKGROUND

Behçet is a systemic inflammatory disease, with global distribution, affecting all ages. The predominant clinical phenotype includes painful ulcers that compromise the oral and genital mucosa, as well as uveitis with a high risk of blindness. It may present as vasculitis affecting different vessel sizes, with complex manifestations in the central nervous system: venous sinus thrombosis, diffuse parenchymal inflammation, cerebral pseudotumor, and optic neuritis. Skin, joints and gastrointestinal tract may also be affected.

The etiology of the disease is still unknown, but there is a genetic association with HLA-B51. The environment may contribute through infectious agents that may induce an attack on the innate immune system in predisposed patients.

CASE REPORT

Case 1 - A 15-year-old male patient consulted in the pediatric emergency room (PER) for panuveitis. After admission, he presented fever, seizures, numbness, oral and genital ulcers and was transferred to the intensive care unit. Bacterial meningitis and brain tumor were discarted. Magnetic resonance imaging (MRI) of cranium evidenced extensive inflammatory lesions involving the left internal capsule and thalamus. The patient received pulse therapy with Methylprednisolone (MP) and Cyclophosphamide (CYC), followed by induction scheme with CYC with excellent clinical response.

Case 2 - Female 12 years old, also consulted on PER due to recurrent oral and genital ulcers, associated with recurrent headache without response to common analgesics. Clinical examination without motor or sensitive changes, MRI of cranium showed active vasculitis involving the left midbrain-pontine transition. Currently in an induction scheme with CYC and MP. Performed MRI after 3 months of therapy, absence of vasculitis.

CONCLUSION

Although it is not known the real prevalence of Behçet's disease in Brazil, it should be part of the differential diagnoses of recurrent and complex childhood aphthosis and panuveitis. Central nervous system manifestations may have a catastrophic evolution and high morbidity in the absence of adequate treatment. Corticosteroid and CYC therapy in these cases demonstrated efficacy for clinical induction, with no severe adverse events observed.