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PEDIATRIC NEURO-BEHÇET'S DISEASE: A RARE, SEVERE AND INCAPACITATING ILLNESS.

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BACKGROUND

Pediatric Behçet disease(BD) has been rarely reported in children and adolescents, particularly associated with central or peripheral nervous system involvements. The objective of the present study, therefore, was to assess clinical manifestations, laboratory abnormalities and outcomes of pediatric neuro-Behçet's disease(NBD).

MATERIALS AND METHODS

A retrospective study evaluated all patients followed in a Pediatric Rheumatology Unit of São Paulo state, Brazil, during the last 35 years(1983-2018). BD was diagnosed by International Study Group-ISG criteria and NBD by International Consensus Recommendation-ICR criteria was based on cerebrospinal fluid-CSF and/or magnetic resonance imaging-MRI abnormalities, and classified as definitive or probable.

RESULTS

First, 43/6,213(0.7%) patients were selected according to recurrent oral ulceration at least three times in one year. BD was diagnosed in 6/6,213 (0.1%). Definitive NBD was diagnosed in 2/6,213(0.03%) and probable NB in 3/6,213(0.04%). All five patients were male. The median age of first neurological manifestation was 9 (8-13) years. The most recognized neurological syndromes were cerebral (n=4) and acute meningeal syndrome (n=4). Parenchymal lesions on MRI were detected in 1 patient, non-parenchymal involvement in 2 and mixed parenchymal/non-parenchymal in 2. The most used treatments were glucorticosteroid (n=5), immunosuppressive (n=4) and biological agents (n=1). Regarding, outcomes incapacitating manifestations (hemiparesis) was observed 3/5 and 2/3 had chronic and recurrent meningitis and headache. None of them died.

CONCLUSION

BD and NBD were rarely observed in a tertiary pediatric rheumatology center. NBD was a severe illness characterized by incapacitating or chronic/recurrent manifestations.