HANDL SYNDROME IN PEDIATRIC AGE

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Introduction

Objectives: The syndrome of transient headache and neurologic deficits with cerebrospinal fluid lymphocytosis (HaNDL) is a rare syndrome of unclear pathogenesis characterized by one or more episodes of severe headache, transient neurologic deficits and lymphocytic pleocytosis in the cerebrospinal fluid, seldom reported in paediatric age. In most cases it is a benign and self limited disorder, although it may mimic various serious, including life-threatening, diseases, such as stroke and meningoencephalitis, which is why vigorous tests should be sought before this diagnosis of exclusion can be reached.

Methods

We report three cases of HaNDL occurred in 2 boys (14 years and 10 years old) and in a 17 years old girl.

Results

Each patient presented with headache, altered conscious state and papilledema associated with different neurological symptoms such as dysarthria, hemiplegia, pernicious vomiting, ideomotor slowing and psychomotorr agitation. None of them had fever and there was no evidence of meningeal irritation.
They received Ceftriaxone, Aciclovir, and Dexamethasone for possible encephalitis and/ or autoimmune disorders. Clinical manifestations were compatible with a variety of disorders including structural brain lesions, meningitis, seizures, autoimmune, vasculitic and paraneoplastic disorders. We performed neuroimaging examinations (CT scan and MRI of the brain), EEG and serum/CSF studies for infectious, autoimmune and vasculitic diseases. All of these aetiologies were ruled out. In one case, a complete tox screen was added and it resulted negative. The laboratory finding common to all three cases was a clear CSF lymphocytic pleocytosis and an elevated opening pressure during lumbar puncture. The intracranial hypertension treated in all three cases with acetazolamide per os with complete remission. In one case, it was necessary the admission in the intensive care unit because of the worsening of psychomotorr agitation of the patient, requiring sedation and endotracheal intubation. All three patients recovered without any neurological sequelae during the follow up

Conclusions

Each patient presented with headache, altered conscious state and papilledema associated with different neurological symptoms such as dysarthria, hemiplegia, pernicious vomiting, ideomotor slowing and psychomotorr agitation. None of them had fever and there was no evidence of meningeal irritation.
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References

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