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Review article

Acute hemicerebellitis in children: Case report and review of literature

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ABSTRACT

Acute hemicerebellitis in childhood is an extremely rare unilateral presentation of cerebellitis mimicking a tumour. Its aetiology is unknown, although an inflammatory or postinfectious origin is presumed. Its clinical outcome is generally good and a self-limited evolution, in the absence of specific treatment, is usually expected. MRI findings can be misunderstood leading to erroneous diagnosis and invasive treatments. Clinical improvement and regression of the pathological findings in serial MRI will help differentiate acute hemicerebellitis from a neoplastic process. Surgical procedures should be performed only in case of clinical deterioration. We present a case of pseudotumoral hemicerebellitis in an eight-year-old girl, presenting with severe headache. This paper provides a review on hemicerebellitis and highlights the clinical, diagnostic, therapeutic features and outcome of this entity.

Contents

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1. Introduction

Acute hemicerebellitis (AHC) is an extremely rare disorder in childhood. Its aetiology is unknown, although an inflammatory or postinfectious origin is presumed. Most cases reported had a good clinical outcome and a self-limited evolution, in the absence of specific treatment, is usually expected. Magnetic resonance imaging (MRI) can be misunderstood leading to erroneous diagnosis and invasive treatments. Our aim was to characterize the clinical picture, aetiology and prognosis of hemicerebellitis, through the report of a new case and the review of literature. We accessed articles on hemicerebellitis, using Pubmed, covering from January 1995 until October 2012. Search terms used were "cerebellitis", "hemicerebellitis", "paediatrics" and "children".

2. Case report

An eight-year-old girl was presented to our service for severe headache for the past eleven days. She reported oppressive fronto-occipital and neck pain of gradual onset, which increased in intensity and became continuous for the last 24 h prior to admission to our unit. Pain limited her daily activities, did not improve with analgesics, woke her up at night and was exacerbated by the Valsalva manoeuvre. Nausea, vomiting, phonophobia, photophobia, fever, redness, tearing and rhinorrhoea were not present. There was no recent viral illness, vaccination or toxic exposure. She was not on any medication. She had no similar episodes before. Her past medical and family history was otherwise unremarkable. No abnormalities were found at physical examination, including cerebellar function and meningism.

Standard cranial computed tomography (CT) revealed a slight supratentorial ventriculomegaly and a fourth ventricle hardly seen. MRI showed high-intensity areas on T2-weighted images in the right cerebellar hemisphere (RCH), compatible with diffuse oedema, producing a mild mass effect over the fourth ventricle, tonsillar herniation and supratentorial hydrocephalus (Fig. 1a,b). There was no involvement of ipsilateral white matter, vermis or contralateral cerebellar hemisphere. Gadolinium-enhanced T1-weighted images demonstrated leptomeningeal enhancement along the cerebellar folia. No perfusion or spectroscopy sequences were done. Lumbar puncture was not performed because of risk of herniation. Regular blood tests were normal. Serological tests for *Epstein–Barr virus*, *human herpes virus* types 1, 2, 6 and 8, *varicella zoster virus*, human immunodeficiency virus, *rubella virus*, *parvovirus B19*, measles virus, mumps virus, *Toxoplasma gondii*, *Brucella melitensis*, *Treponema pallidum*, *Rickettsia conorii*, *Borrelia burgdoferi*, *Mycoplasma pneumonie* and *Bartonella henselae* in serum were all negative. No cultures or other microbiological tests were performed. Tests for autoimmunity were normal, including normal serum concentrations of rheumatoid factor and complement and of antinuclear, antiphospholipid, antithyroid, anti-transglutaminase, antigliadin, anti-double-stranded DNA and anti-ENA (extractable nuclear antigens) antibodies.

The rapid onset of symptoms together with the foliar enhancing pattern and the absence of a well-delimited mass, suggested an AHC of inflammatory origin rather than a tumour. The present foliar enhancing pattern has been advocated as specific of cerebellitis.[1] She was put on treatment with acyclovir (80 mg/kg/day), cefotaxime (150 mg/kg/day) and dexamethasone (1 mg/kg/day). Symptoms disappeared after the first day of treatment. Antibiotic and acyclovir were suspended after 48 h due to a positive clinical evolution and negative results on the microbiological tests. Corticoids were continued for eight days. She was then discharged from hospital and has remained asymptomatic to date. The first follow-up visit, six months after discharge, revealed a normal physical examination. Follow-up MRI at the time showed global reduction of the RCH volume, with a slight increase in the signal of the RCH cortex in T2-weighted images, compatible with atrophy and residual gliosis (Fig. 2). At the last follow-up appointment, sixteen months after the episode, the neurological examination remained normal. The satisfactory clinical evolution and the results on the follow-up MRI confirmed the previously suspected inflammatory process.

3. Discussion

Acute cerebellar ataxia (ACA) is a relatively common cause of presentation to the paediatric emergency room or a child neurologist.[2] Acute cerebellitis (AC) is a rare syndrome characterized by cerebellar symptoms and MRI changes primarily confined to the cerebellum.[3–5] Both conditions occur mostly in children and represent a process characterized by parainfectious, postinfectious, or postvaccination cerebellar inflammation; hence, some groups have claimed an overlap between these entities.[6] AC is usually considered a benign condition.[7,8] Nevertheless, severe cases presenting with alteration of consciousness, focal neurological signs, hydrocephalus, herniation or even fulminant course have been reported.[6,9–13] Cognitive disorders in spatial visualization ability, language skills and concentration have been reported as long-term sequelae of children with cerebellitis.[6,14,15]

Acute hemicerebellitis (AHC) is an extremely rare entity that could represent a variant of AC limited to one cerebellar hemisphere. There is no current explanation for such a restricted unilateral affection; we suggest that an impaired cerebral blood flow or an anatomic variant of the posterior circulation system could be responsible for that. Another group raises the possibility of a mild subclinical involvement of the contralateral cerebellar hemisphere.[16] Twenty-one