Neuroimaging of Infectious and Inflammatory Diseases of the Pediatric Cerebellum and Brainstem

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KEYWORDS

• Ataxia • Cerebellitis • Cerebellar infections • Rhombencephalitis

KEY POINTS

- Infectious and inflammatory diseases of the cerebellum are rare in the pediatric age group; most patients are previously healthy preschool children presenting with an acute onset of cerebellar ataxia.
- Acute postinfectious cerebellar ataxia (a benign, self-limiting disease) and acute cerebellitis (a serious, potentially life-threatening disease) may represent 2 ends of a spectrum of viral involvement of the cerebellum, typically in the course of varicella infection.
- The cerebellum may be involved in acute disseminated encephalomyelitis, multiple sclerosis, and acute necrotizing encephalopathy, typically in the context of widespread central nervous system disease.
- Cerebellar abscesses are mostly otogenic in the pediatric age group; an associated dural sinus thrombophlebitis must actively be excluded.
- Congenital infections, mainly cytomegalovirus, may impair cerebellar development/maturation, resulting in cerebellar hypoplasia with abnormal foliation.

INTRODUCTION

The rhombencephalon, or hindbrain, is composed of the pons, cerebellum, and medulla oblongata. The term derives from the Greek rhombos, meaning a lozenge-shaped figure, whereas enkephalos means the brain. During early embryogenesis, the primary rhombencephalic vesicle divides into 2 secondary vesicles: the metencephalon (eventually forming the pons and cerebellum) and the myelencephalon (eventually forming the medulla oblongata). Rhombencephalic involvement by infectious-inflammatory conditions is rare in children. Most affected patients present acutely with ataxia, which is defined as the inability to coordinate muscle activity in the execution of voluntary movements. These children are mostly young (typically preschool) and previously healthy, although preexisting prodromal symptoms such as fever or rash, or chronic medical disorders, are sometimes recorded. Clinically, patients

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complain with broad-based gait, dysmetria, dysdiadochokinesia, intention tremor, dysarthria, and sometimes nystagmus, all of which progress rapidly to a nadir, which is reached within hours to 1 to 2 days of the onset.[1]

In this clinical context, the term cerebellitis is often used loosely by clinicians to indicate that the acute onset of cerebellar ataxia likely results from primary infection or a postinfectious immune-mediated process involving the cerebellum. However, within this broad category several conditions with often different presentation, outcome, and therapeutic implications are included. The term rhombencephalitis (RE) is also often used loosely, mainly to refer to inflammatory diseases that involve primarily the brainstem; the term brainstem encephalitis is often used interchangeably with RE. The differential diagnosis of patients presenting in the emergency department with acute cerebellar ataxia includes a large group of entities, such as vascular, traumatic, metabolic, and neoplastic diseases next to infectious-inflammatory causes.[1] More specific terminology and agreement about the usage of nomenclature is therefore desirable.

The term acute cerebellitis should be used to refer to an infection that directly affects the cerebellum, either unilaterally or bilaterally, often with abnormal magnetic resonance (MR) imaging findings and with symptoms and signs beyond those of a pure cerebellar syndrome. In contrast, the term acute postinfectious cerebellar ataxia (APCA) should be used to indicate a postinfectious (mostly viral) dysfunction, usually presenting with a pure pancerebellar syndrome and with an initially normal MR imaging scan.[2] Despite these attempts at a correct definition, there is considerable overlap between these entities, which may represent a spectrum of disorders ranging from benign, self-limiting disease to marked swelling of the cerebellum and hydrocephalus. Confusion is also generated by other more widespread central nervous system (CNS) inflammatory disorders, such as acute disseminated encephalomyelitis (ADEM) or multiple sclerosis, that can present with predominantly cerebellar clinical findings but with lesions on neuroimaging that are usually widespread.[3] This article reviews the most common infectious and inflammatory conditions involving the cerebellum and brainstem, with a focus on the role of neuroimaging studies, particularly MR imaging scans, in the work-up of affected children.

NEUROIMAGING: GENERAL PRINCIPLES

Children presenting in the emergency department with an acute onset of cerebellar ataxia are typically referred for urgent imaging studies, which may include a computerized tomography (CT) or MR imaging scan depending on several considerations, including the clinical severity, the need for sedation, and organizational issues at the individual facility. Often, the range of differential diagnosis at this initial stage is wide, and includes tumors involving the brainstem or cerebellum and potentially causing hydrocephalus, metabolic disorders, cerebrovascular causes, trauma, and intoxications; thus, the inclusion of neuroimaging studies in the initial work-up of these patients is fully justified.[4,5] Furthermore, the widespread use of varicella and mumps vaccinations has significantly reduced the number of the pediatric cases of APCA.[6] Patients presenting with acute cerebellar ataxia in the postvaccination era may be presumed to harbor significant disorder in a higher proportion of cases. However, children less than 3 years of age and with duration of ataxia of less than 3 days may represent a low-risk group in which neuroimaging can be deferred if contingent on close clinical follow-up and reassessment.[7]

Cranial ultrasonography provides an excellent bedside examination in premature and term newborns and small infants; however, the exploration of the brain is limited to the first months of life in relation to the patency of the acoustic windows, represented by the cranial fontanelles. A recent study in preterm infants showed that the routine use of ultrasonography through the mastoid fontanelle (MF) may allow a better identification of cerebellar hemorrhages than through the anterior fontanelle[8]; this can easily be explained by the closer and better visualization of posterior fossa structures, both on coronal and axial planes, that can be obtained through the MF, particularly in extremely immature newborns. However, the exploration of the cerebellum remains challenging, and there are differences in the diagnosis and interpretation of cranial ultrasonography examinations according to the available ultrasonography technology and expertise.[9]

The authors prefer MR imaging to CT because of its intrinsically higher contrast resolution and sensitivity, particularly for lesions affecting the posterior fossa, a notoriously difficult region for CT despite newer technological advances,[10] as well as for inherent radiation issues. However, when MR imaging is unavailable or unfeasible, CT remains an important imaging modality. A helical acquisition with reformatted images in 3 planes should be obtained, and the whole brain should be studied including the craniocervical junction and cervical spine until C3. Iodinated contrast material administration is not routine, and should be performed only when there is concern for