Acute disseminated encephalomyelitis
Published on 24.06.2005

DOI: 10.1594/EURORAD/CASE.3395
ISSN: 1563-4086
Section: Paediatric radiology
Case Type: Clinical Cases
Authors: Rea D, Ryan S
Patient: 5 years, female

Clinical History:
A five-year-old girl presented with a left facial nerve palsy, left hemiplegia, seizures and a recent respiratory tract infection. An MRI was done, which demonstrated multiple foci of asymmetric hyperintensity involving the white matter, thalami and the brainstem.

Imaging Findings:
A five-year-old girl presented with an acute onset of left facial nerve palsy and a moderate left hemiplegia, associated with generalised seizure activity. There was a history of viral respiratory tract infection in the week prior to the onset of neurological symptoms. A contrast enhanced CT scan of the brain was initially performed, which demonstrated multiple asymmetric areas of reduced attenuation. An MRI of the brain was done, which showed multiple foci of asymmetric hyperintensity on T2 and FLAIR sequences involving the peripheral white matter–grey matter junction, thalami and extending into the brainstem. These lesions demonstrated an enhancement following an intravenous injection of gadolinium. Following treatment with high dose of corticosteroids, there was a resolution of the clinical findings with almost complete resolution of the multiple white matter abnormalities on follow-up MR imaging.

Discussion:
Acute disseminated encephalomyelitis (ADEM) is typically a monophasic autoimmune mediated white matter demyelination of the brain and/or the spinal cord, usually followed by remyelination. Cortical and deep grey matter can be involved but to a lesser extent than white matter. The ADEM often follows recent viral infection, vaccination, respiratory infection or exanthematous disease of childhood. The onset of the neurological symptoms is typically 5–14 days following the onset of the acute viral illness. It is proposed that the underlying aetiology relates to an autoimmune cell mediated cross reaction response against myelin sensitised by viral protein. The ADEM is seen predominantly in the paediatric age group and is rarely seen in adults. The peak onset occurs between 3 and 5 years but it can occur at any age. The clinical manifestations of ADEM are often preceded by a prodromal phase characterised by viral features including fever, malaise and myalgia. Subsequent neurological symptoms tend to be multifocal and may initially present with headache fever, or drowsiness. Cranial nerve palsies, seizures, hemiparesis and altered levels of consciousness have also been described. The symptoms may be similar to those seen in a single attack of multiple sclerosis. An examination of the CSF frequently demonstrates the occurrence of leucocytosis and an elevated protein level and the absence of oligoclonal bands. A computed tomography (CT) scanning may be normal in up to 40% of the patients with ADEM. Magnetic resonance imaging (MRI) is the modality of choice for diagnosis and during follow-up. T2-weighted sequences demonstrate the typical multiple bilateral asymmetric hyperintense lesions and FLAIR sequences allow the abnormalities be best appreciated. The white matter abnormalities can involve the white matter–grey matter junction and the brainstem or posterior fossa. In contradistinction to multiple sclerosis, abnormalities at the callososeptal interface are not commonly seen.
Tumefactive hyperintense mass lesions are reported and these appear to have less mass effect than would be expected for their size. Post-gadolinium imaging usually shows the punctate ring or nodular enhancing pattern. The absence of enhancement does not exclude ADEM as a diagnosis. The initial imaging findings may be similar to those seen in multiple sclerosis, therefore, repeat follow-up MR imaging should be performed. No new lesions should appear after approximately 6 months after the onset of ADEM. Acute haemorrhagic leucoencephalitis (Hurst’s Disease) is a rare aggressive form of ADEM accounting for 2% of all cases. It is a diffuse multifocal perivascular condition associated with demyelination and haemorrhage confined to the cerebral white matter with sparing of the subcortical U-fibres. Bilateral striatal necrosis is a common imaging feature. Death usually occurs within six days of onset of symptoms due to cerebral oedema and herniation. Survivors of this form of ADEM tend to have severe neurological deficits. Gadolinium enhancement may be poor and MR may not always demonstrate the associated cerebral haemorrhages. The prognosis for ADEM is variable. Approximately 50%–60% of the patients make a complete recovery in one month. Between 20% and 30% suffer some neurological sequelae (most commonly seizures) as a result of the occurrence of the disease. Mortality varies between 10% and 30%. The therapy is supportive, including ventilation if required. The immunomodulatory agents, such as steroids, immunoglobulins and plasmapheresis, may have a role, although their benefit remains largely unproven.

**Differential Diagnosis List:** Acute disseminated encephalomyelitis.

**Final Diagnosis:** Acute disseminated encephalomyelitis.

**References:**


