Official Journal of the European Paediatric Neurology Society

Case study

Acute demyelinating events with rhombencephalitis: A high-risk subgroup in children

S. Tirupathi[a,b,*], N. Lynch[a], E. Phelan[c], J. Mc Menamin[a,b], D. Webb[a]

[a]Department of Neurosciences, Our Lady's Hospital, Dublin, Ireland
[b]Department of Paediatrics, Royal College of Surgeons, Ireland
[c]Department of Radiology, Our Lady's Hospital, Dublin, Ireland

ARTICLE INFO

Article history:
Received 18 April 2007
Received in revised form
18 June 2007
Accepted 17 July 2007

Keywords:
ADEM
CIS
Mass effect
Posterior fossa
Rhombencephalitis

ABSTRACT

Acute, acquired demyelination of central nervous system in childhood leads to a variety of clinical phenotypes dependant on the site of demyelination and presence of encephalopathy. Posterior fossa involvement is seen in over third of cases in childhood. We report on four children who had cerebellar involvement with significant mass effect in posterior fossa on CT and MRI brain. This subgroup of children have significant cerebellar and brainstem swelling (rhombencephalitis) and is difficult to distinguish entirely on clinical grounds from other children with acute demyelinating events at presentation.

1. Introduction

Acute, acquired demyelination of central nervous system in childhood leads to a variety of clinical phenotypes dependant on the site of demyelination and presence of encephalopathy. Acute disseminating encephalomyelitis (ADEM) is a first clinical event with presumed inflammatory or demyelinating cause with acute or subacute onset that affects multifocal areas of the CNS. The clinical presentation must be polysymptomatic and must include encephalopathy (confusion, excessive irritability, lethargy, coma) and neuroimaging shows focal or multifocal lesion(s).[1] It commonly affects subcortical white matter, most often bilaterally and asymmetrically in the frontal and parietal lobes.[2,3] Posterior fossa involvement is seen in over a third of cases in childhood.[2,3] Clinically isolated syndrome (CIS) is a first acute clinical episode of CNS symptoms with presumed inflammatory cause for which there is no prior demyelinating event and can be monofocal or multifocal but usually does not include encephalopathy (except in brainstem syndromes).[1]

The aim of this report is to increase awareness of a subgroup of patients with demyelinating disorders with predominant involvement of the posterior fossa that is potentially life threatening.

1.1. *Case reports*

Among 32 children presenting with post-infectious CNS disorders to our service between 1999 and 2004, 12 had evidence of demyelination on neuroimaging. Eight were

*Corresponding author. Department of Neurosciences, Our Lady's Hospital, Crumlin, Dublin 12, Ireland. Tel.: +353 14096413; fax: +353 14096887.
E-mail address: sanshek123@yahoo.com (S. Tirupathi).
1090-3798/$ - see front matter © 2007 European Paediatric Neurology Society. Published by Elsevier Ltd. All rights reserved.
doi:10.1016/j.ejpn.2007.07.004

Table 1 – Summary of findings in the four children

Patient details	Clinical presentation	Neurological findings	Emergency neuroimaging	Course and EDSS
Case 1: Boy aged 2.5 years	URTI for 4 days. Admitted with abdominal pain, irritability and lethargy over 24 h, followed by generalized seizures.	GCS 7 (E2, M3, V2). Focal right sided motor seizures. Generalized hypertonia+extensor plantars.	CT: Cisternal effacement and fourth ventricle compression. Loss of grey white differentiation in the occipito-parietal area.	Diagnosis: ADEM Nearly full recovery within a week. EDSS at 3 years: 2
Case 2: Girl aged 7 years	Headache, lethargy, and vomiting for 7 days followed by unsteadiness and slurred speech.	Truncal ataxia, dysmetria and intention tremor. Normal tone and reflexes.	CT: Cisternal effacement and fourth ventricle compression.	Diagnosis: CIS Full recovery within 6 weeks. EDSS at 7.5 years: 0
Case 3: Boy aged 10 years	Flu-like illness, with headache, photophobia, lethargy and vomiting for 3 days. Found in bed in a seizure.	GCS 5 (E1, V2, M2). Intermittent unilateral pupillary dilatation. Tonic downward deviation of the eyes. Decerebrate, hyper-reflexic, extensor plantars.	CT: Cisternal effacement and fourth ventricle compression with secondary hydrocephalus.	Diagnosis: ADEM Extraventricular drain inserted. Mild residual limb ataxia. EDSS at 10.5 years: 2
Case 4: Girl aged 11 years	Fever, vomiting, lethargy and headache for 4 days. Admitted with irritability and confusion and developed bulbar signs with decerebrate posturing.	GCS 5 (E2, V1, M2). PEARL. Bulbar paresis. Limb hypotonia and extensor plantars.	CT: Fig. 2 Cisternal effacement and fourth ventricle compression.	Diagnosis: ADEM Ventilated. Initial Spastic quadriparesis-recovered over 4 weeks. EDSS at 11.5 years: 1

EDSS: Expanded Disability Status Score; GCS: Glasgow Coma Scale.

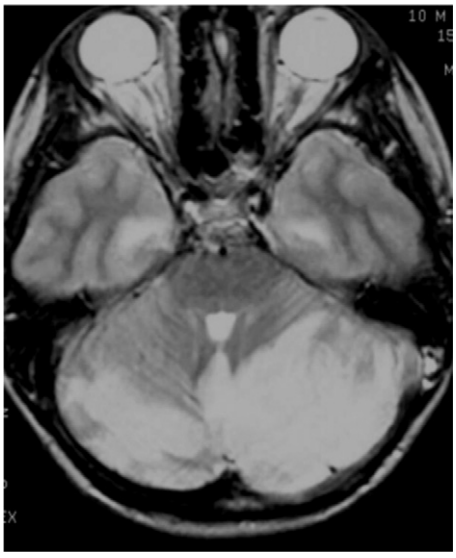

Fig. 1 – Focal areas of swelling in cerebrum and bilateral cerebellar swelling (Case 3).

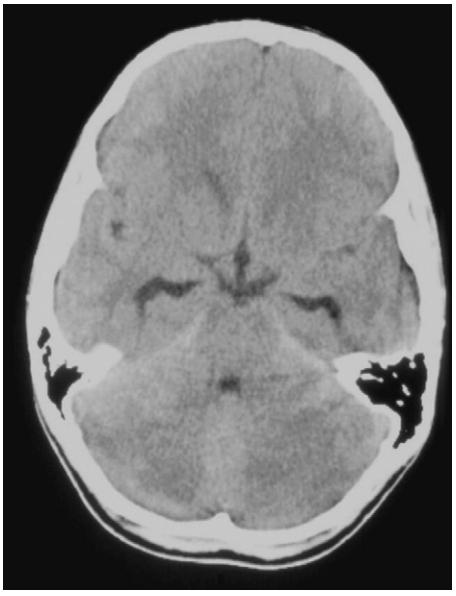

Fig. 2 – CT: Cerebellar swelling with effacement of the basal cisterns and small fourth ventricle (Case 4).

diagnosed with ADEM, three with CIS and one with recurrent ADEM. Four of these 12 children had cerebellar involvement with significant mass effect in the posterior fossa on CT and MRI brain. Three had encephalopathy with demyelinating lesions (ADEM) and one had demyelinating lesions confined to the cerebellum (CIS).

Clinical details of the four patients are summarized in Table 1. The four children were previously normal and had no family history of demyelinating disease. The children were aged 2, 7, 10 and 11 years at presentation. Three required