ELSEVIER

Brain & Development xxx (2018) xxx–xxx

BRAIN & DEVELOPMENT
Official Journal of the Japanese Society of Child Neurology

www.elsevier.com/locate/braindev

Case Report

Bickerstaff's brainstem encephalitis with overlapping Guillain-Barre' syndrome: Usefulness of sequential nerve conduction studies

Choong Yi Fong [a,*], Hnin Wint Wint Aung [a,b], Arie Khairani [a,c], Chin Seng Gan [d], Nortina Shahrizaila [e], Khean Jin Goh [e]

[a] *Division of Paediatric Neurology, Department of Paediatrics, Faculty of Medicine, University of Malaya, Kuala Lumpur, Malaysia*
[b] *Paediatric Neurology Unit, Yangon Children Hospital, Yangon, Myanmar*
[c] *Division of Paediatric Neurology, National Brain Centre, East Jakarta, Jakarta, Indonesia*
[d] *Division of Paediatric Intensive Care, Department of Paediatrics, Faculty of Medicine, University of Malaya, Kuala Lumpur, Malaysia*
[e] *Division of Neurology, Faculty of Medicine, University of Malaya, Kuala Lumpur, Malaysia*

Received 21 November 2017; received in revised form 29 January 2018; accepted 1 February 2018

Abstract

Bickerstaff's brainstem encephalitis (BBE) is a rare immune-mediated disorder characterized by ophthalmoplegia, ataxia and disturbance of consciousness, which may overlap with Guillain-Barré syndrome (GBS) if there is additional limb weakness. We report a 7-month-old boy presented with ophthalmoplegia followed by a rapidly ascending paralysis of all four limbs and disturbance of consciousness. The initial impression was BBE with overlapping GBS. This was supported by sequential nerve conduction study (NCS) findings compatible with an acute inflammatory demyelinating polyneuropathy (AIDP). He received intravenous pulse methylprednisolone, intravenous immunoglobulin and plasmapharesis with complete clinical recovery after 6 weeks of illness and improved NCS findings from week 16. This is the first case of paediatric BBE with overlapping GBS with an AIDP subtype of GBS. It expands the clinical spectrum of this condition in children. Our case highlights the importance of sequential NCS in paediatric BBE with overlapping GBS for accurate electrophysiological diagnosis and prognosis particularly if the first NCS findings are not informative.

Keywords: Bickerstaff's brainstem encephalitis; Gullain-Barre' syndrome; Acute inflammatory demyelinating polyneuropathy; Nerve conduction study

1. Introduction

Bickerstaff's brainstem encephalitis (BBE) is a rare immune-mediated disorder in children. BBE is characterized by acute onset of ophthalmoplegia, ataxia, and disturbance of consciousness [1]. Guillain-Barre' syndrome (GBS) is an immune-mediated polyradiculoneuropathy characterized by a rapidly progressive symmetrical ascending weakness [2]. It is recognized that both these disorders share many common features, in particular the antecedent infection, albuminocytological dissociation and the presence of antiganglioside antibodies [3]. Co-existence of the two syndromes results

* Corresponding author at: Division of Paediatric Neurology, Department of Paediatrics, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia.
E-mail address: cyfong@ummc.edu.my (C.Y. Fong).

https://doi.org/10.1016/j.braindev.2018.02.001
0387-7604/

in limb weakness; in addition to all the other clinical features of BBE [4]. The electrophysiological findings also play an important role in making the diagnosis of BBE with overlapping GBS.

Paediatric BBE with overlapping GBS is rare. There are only 2 cases reported to date [5,6]. We describe a 7-month-old boy with a diagnosis of BBE with overlapping GBS and sequential nerve conduction study (NCS) findings support the electrodiagnosis of acute inflammatory demyelinating polyneuropathy (AIDP). This is the first case report of BBE and overlapping GBS with sequential NCS findings confirming an AIDP subtype of GBS. Written informed consent has been obtained from parents to publish this case report. No institutional ethical approval was required for this case report.

2. Case report

A 7-month-old boy presented to our hospital because of respiratory failure and progressive ascending paralysis. He had a 1 week prior history of diarrhoea and coryzal symptoms. On day 1 of illness, he developed bilateral ophthalmoplegia associated with weakness of both lower limbs which progressed to involve both upper limbs and he developed respiratory failure requiring mechanical ventilation. On day 5 of illness, he developed impaired brainstem reflexes (absent corneal, oculocephalic, and gag reflexes). On day 6 of illness, his muscle power Medical Research Council (MRC) deteriorated to grade 0 for all limbs with areflexia and by day 10 of illness he became encephalopathic and developed autonomic instability with hypertension and tachycardia. Electroencephalogram (EEG) on day 10 showed moderate encephalopathy.

Brain and spine magnetic resonance imaging (MRI) on day 5 of illness showed lumbar spine nerve root enhancement extending to the cauda equina with no abnormal signal in the brain. Cerebrospinal fluid (CSF) on day 5 of illness showed albuminocytological dissociation [cell count: 0/μL with elevated protein: 0.52 g/L (normal range: 0.08–0.32 g/L)]. Serum and CSF antiganglioside antibodies (Ig G and Ig M antibodies of GM 1, GM 2, GM 3, GM 4, GD 1a, GD 1b, GD 2, GD 3, GT 1a, GT 1b and GQ 1b) were negative. Stool for *Campylobacter jejuni* was negative. The patient was treated with high dose intravenous methylprednisolone (30 mg/kg/day) from day 5 to 7 of illness and intravenous immunoglobulin (1g/kg/day) from day 5 to 6 of illness. Four cycles of alternate day plasmapharesis was performed from day 19 of illness. He showed gradual clinical improvement with resolution of hypertension and tachycardia, and improvement in muscle power following the second cycle of plasmapharesis. He was ventilated for 21 days. From day 26 of illness he showed significant clinical recovery with muscle power of MRC grade 3 bilaterally and made a complete clinical recovery from week 6 of illness.

Sequential NCS was performed over the right upper and right lower limbs at day 6, week 4, week 6, week 10, week 16 and week 29 of illness respectively (Table 1 and Fig. 1). Initial NCS on day 6 showed absent median, ulnar, posterior tibial and common peroneal compound muscle action potentials (CMAP) and absent median, ulnar, radial and sural sensory nerve action potentials (SNAP). By week 4, median and tibial CMAPS were recordable but with prolonged distal motor latencies (DML), small CMAP amplitudes and markedly slow median motor nerve conduction velocity (MNCV) fulfilling the electrodiagnostic criteria for AIDP [7,8]. The ulnar and peroneal nerve CMAPs remained absent. At week 6, all four CMAPs were recordable with definite demyelination features with slow MNCV, prolonged DML and ulnar motor conduction block in the forearm. F-waves and SNAPs remained absent. By week 10, CMAP amplitudes had improved substantially except the peroneal nerve and the ulnar motor conduction block had resolved. F-waves with prolonged latencies were recordable in all motor nerves but MNCV remained slow in all nerves. NCS from week 16 onwards, showed continued improvement of all motor nerves parameters and a subsequent return to normal limits. Of the initially absent SNAPs, the sural nerve was first to recover at week 10 but median and ulnar SNAP were recordable on later NCS.

3. Discussion

BBE with overlapping GBS is diagnosed in patients with subacute onset of progressive external ophthalmoplegia, ataxia, and impairment of consciousness with additional limb weakness (MRC power <3) [4]. In the current case, a diagnosis of BBE with overlapping GBS was made because our patient fulfilled the clinical criteria stated above, together with the CSF, NCS and MRI spine findings typical of GBS. There have been numerous reports of BBE overlapping with GBS in the adult population. In a large series of adult BBE cases, 60% of patients were associated with GBS [1]. In another case series only 2 of 14 adult BBE with overlapping GBS patients showed features of demyelination on NCS and the majority of reports show that patients had axonal degeneration subtype as the predominant feature with more severe cases demonstrating inexcitable nerves [4,7].

In contrast, there are only two published paediatric cases of BBE with limb weakness. Table 2 shows a comparison of clinical features between our case with the reported paediatric cases [5,6]. None of the paediatric BBE with overlapping GBS cases had sequential NCS to confirm their electrodiagnosis. Our case is the youngest reported paediatric case and the first to show clear features of AIDP on sequential NCS correlating with the faster and complete clinical recovery when compared

Please cite this article in press as: Fong CY et al. Bickerstaff's brainstem encephalitis with overlapping Guillain-Barre' syndrome: Usefulness of sequential nerve conduction studies. Brain Dev (2018), https://doi.org/10.1016/j.braindev.2018.02.001