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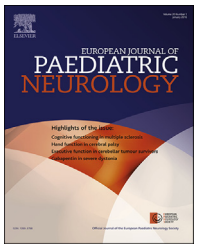

Case Study

CMV-associated axonal sensory-motor Guillain–Barré syndrome in a child: Case report and review of the literature

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ABSTRACT

Background: Guillain–Barré syndrome is the most frequent cause of flaccid paresis in Western countries. Moreover, CMV infection is the most common antecedent viral infection in adult patients and the presence of specific IGM antiganglioside antibodies is often identified. Instead, Guillain–Barré syndrome following CMV infections is rarely reported in childhood and often presents severe symptoms at onset and longer recovery times.
Material and methods: One year of clinical, electrophysiological and serological follow-up of a 9-year old child with axonal sensory-motor Guillain–Barré syndrome following CMV infection is reported. Moreover, the literature data on paediatric sensory-motor axonal GBS and GBS secondary to CMV infection and antiganglioside antibodies are reviewed.
Results: Our patient presented with paraesthesias and a pattern of weakness showing proximal predominance and affecting the upper limbs more than the lower limbs. At nadir, unilateral facial palsy was also present and he was unable to walk. Electroneurography showed motor-sensory axonal damage. Both anti-CMV and anti-GM2 IgM were positive. After early treatment with IVIG and IV methylprednisolone the patient recovered deambulation. Six months later, his neurological examination was normal and electroneurography showed normal data.
Conclusion: The sensory-motor axonal form of Guillain–Barré syndrome following CMV infection may present a good prognosis and a prompt full recovery also in children, if adequate treatment is started in time.

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Table 1 – Paediatric cases of CMV-related Guillain–Barré syndrome.

CMV-related GBS paediatric cases

Number of pts	Age, gender	Prodromes	Cranial nerves involvement	Sensory involvement	Weakness pattern	ANS involvement	Additional clinical features	ENG/EMG diagnosis
n = 1	17 y, F	None	Facial + bulbar palsy	Dysesthesia Hypoesthesia	NR	NR	NR	AIDP
n = 3	13 y, 15 y, 17 y 3F	2 URTI 1 none	NR 3/3	Distal anaesthesia /analgesia: 1/3, symptoms but not signs: 2/3	NR	NR	NR	NR
n = 1	15 y, F	URTI	Mild facial palsy, dysarthria, swallowing dysfunction	No	Limbs weakness (ULs > LLs) (pharyngo-cervicobrachial variant)	No	No	Unclassified (normal motor & sensory NCV, bilateral ulnar & peroneal conduction block)
n = 1	15 y, F	None	Not involved	LLs paraesthesias; symm ↓ vibration, light touch, & proprioception below knees	Distal LLs	No	Pregnant (10th wG)	AIDP

List of abbreviations: AIDP: acute inflammatory demyelinating neuropathy, ANS: autonomous nervous system, CMV: cytomegalovirus, ENG/EMG: electroneurography/electromyography, F: female, GBS: Guillain–Barré syndrome, IVIG: intravenous immunoglobulins, LLs: lower limbs, n: number, NCV: nerve conduction velocity, NR: not reported, pts: patients, Ref: reference, ULs: upper limbs, URTI: upper respiratory trait infection, wG: weeks of gestation, y: years.

1. Introduction

Guillain–Barré syndrome (GBS) is an acute inflammatory polyneuropathy characterised by acute onset and rapid progressive symmetric weakness and areflexia. It has been subdivided into several subtypes although two major forms are recognised: inflammatory demyelinating and acute motor axonal neuropathy. GBS occurring in children usually displays milder degrees of severity,[1] and motor involvement is more often accompanied by sensory symptoms, such as pain or paraesthesias, ataxia or cranial nerves involvement.[2] There is less need for mechanical ventilation (4–15%), and the long-term outcome is generally considered as better than in adulthood.[2] Pathophysiology is post-infectious autoimmune, therefore the onset of acute polyradiculoneuritis is often preceded by an upper respiratory trait or gastrointestinal infection,[3] recognised in up to 75% of paediatric cases.[2]

The aim of this report is to describe a paediatric case of acute motor-sensory axonal neuropathy (AMSAN) secondary to CMV infection with positive anti-GM2 antibodies. Furthermore, literature on paediatric GBS cases of sensory-motor axonal type and of GBS following CMV infection was reviewed.

2. Material and methods

We report one year of clinical, electrophysiological and serological follow-up of a 9-year old child with axonal sensory-motor GBS following CMV infection. Data were collected through revision of clinical charts and neurophysiological and laboratory reports. Furthermore, the scientific literature on paediatric sensory-motor axonal GBS and paediatric GBS secondary to CMV infection, with special reference to antiganglioside antibodies investigations was reviewed. The search was limited to articles in English and it was performed on PubMed database using the Boolean expressions (CMV) OR (cytomegalovirus) AND (Guillain–Barré syndrome) AND (children) OR (paediatric). A second search on PubMed was based on the use of the Boolean expressions (AMSAN) OR (sensory-motor axonal) AND (Guillain–Barré syndrome) AND (children) OR (paediatric). Additional papers were retrieved by manually searching references in selected papers in order to find additional descriptions of CMV-associated or sensory-motor axonal GBS cases. For the CMV-associated cases, articles in which it was not possible to clearly distinguish CMV-positive from CMV-negative cases were excluded, as well as articles in which paediatric cases were not clearly distinguished from adult ones. Only cases exclusively positive for CMV were considered, while cases of possible co-