

Chronic Inflammatory Demyelinating Polyradiculopathy with Multiple Cranial Nerve Palsy: An Atypical Pediatric Case

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Abstract

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is relatively rare in children. Moreover, atypical presentation remains a diagnostic challenge for physician. Early recognition and proper immunomodulatory therapy is important for favorable outcomes with prevention of progressive course. We report a case with atypical CIDP. The aim was to focus on the diagnostic difficulties and the importance of recognizing this treatable condition. [*Journal of National Institute of Neurosciences Bangladesh, 2020;6(1): 64-66*]

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Introduction

Chronic inflammatory demyelinating polyradiculopathy is an auto immune neuropathy usually presented with progressive, proximal and distal symmetric weakness, accompanying with hyporeflexia and sensory symptoms developing over at least 2 month¹. While the incidence among adults is 1.0 to 1.9 per 100,000, the CIDP incidence among patients under age 20 is 0.48 per 100,000². Childhood CIDP is relatively rare³ and the diagnosis is more difficult with atypical clinical or neurophysiologic findings who fail to fulfill the established criteria but potentially treatable and with favorable outcomes. Therefore, early diagnosis and

initial treatment decline demyelination and prevent secondary axonal loss. We report a case of atypical childhood CIDP with multiple cranial nerve palsy.

Case Presentation

A ten years old girl who was the first issue of non-consanguineous parents presented with weakness of all four limbs for three and half (3½) months. Initial concern was sudden weakness in both lower limbs which was symmetrical and progressive. With worsening proximal and distal weakness, increasing difficulties with fine motor coordination, and deterioration in performance of activities of daily living and she became

unable to walk. Along with this she had tingling and numbness over limbs. She had no prior history of gastroenteritis and respiratory tract infection. Last 12 days she experienced double vision with drooping of left eye. There was no swallowing and breathing difficulty, altered mentation, muscle twitching, skin rash and joint pain. She had no history of exposure to toxin or taking any offending drugs. Normal bowel and bladder function was maintained. Her birth and family history was unremarkable for neurological disease. Physical findings revealed a cooperative girl had complete ophthalmoplagia with ptosis on left eye (Left sided III and IV nerve palsy), pupils were mid dilated with sluggish light response, bilateral VI nerve and right VII nerve (LMN) palsy, fundus were normal. She had atrophy of all limb muscles and her muscle power was subnormal (3/5) in the lower limbs and (4/5) in upper limbs. Deep tendon reflexes (DTRs) were absent. There was nothing remarkable regarding sensory function including touch, pain, vibration, and position sense. Cerebellar function was intact. Gait could not be elicited. Her other systems examination was normal. Laboratory tests including complete blood cell count (Hb% 12gm/dl, total count - 9000/cumm, neutrophil (52.0%), platelet (299,000/cumm), serum muscle and liver enzyme levels, thyroid function tests and Vitamin B₁₂ were all normal. On CSF examination, the parameter were cell (8/cumm), lymphocyte (100%) and the high protein level (254 mg/dL). Nerve conduction study (NCS) report had diagnosed as CIDP. She was given IV methylprednisolone 30mg/kg/day for 5 days and oral prednisolone was continued.

Discussion

Chronic inflammatory demyelinating polyneuropathy is an acquired immunological disorder. It is relatively uncommon in children and may cause long term disability if not treated promptly. Several children may have more recurrent relapses than adults, but they mostly respond to treatment and tend to have a more encouraging long-term outcome than adults^{4,5,6}. Several children with CIDP can experience complete remission or stable remaining deficits without the need for additional treatments. Chronic inflammatory demyelinating polyneuropathy (CIDP) in children presents with symmetric, mostly motor neuropathy, evolving over several weeks or months, frequently developing to delay or loss of ambulation. Upper extremity weakness, hand tremor⁷ and ataxia⁸ are also seen in some cases. There is also reduced or absence of deep tendon reflexes⁹. Some studies showed that at least

one-third of pediatric patients may have sensory symptoms such as paresthesias, dysesthesias and large fiber sensory loss³ in absence of distinct biological marker, the diagnosis is based on clinical criteria along with neurophysiologic, neuroimaging, cerebrospinal fluid and nerve biopsy. Systematic diagnostic criteria for childhood chronic inflammatory demyelinating polyneuropathy were devised in 2000 by the European Neuromuscular Consortium¹⁰.

There is wide range of presentation in childhood CIDP. Various patients may show multifocal acquired demyelinating sensory and motor neuropathy (MADSAM). Connolly illustrated two major clinical types of CIDP in children. The first was a monophasic disorder getting maximal weakness over three months and the second was a disorder that have more slowly a relapsing and remitting course¹¹. Monomelic neuropathy is also described as a variation in CIDP. McDonald et al¹² presented a child of nine years' old who showed right upper limb weakness; she was lost to follow-up, but re-presented three years later with extensive progressive weakness in her other limbs. She showed improvement in the right arm after years. Jha et al¹³ reported rare features of CIDP with phrenic nerve involvement and respiratory failure that required mechanical ventilation. Ryan et al¹⁴ describe three in a group of 16 children with mild respiratory symptomatology. One exceptional case was described, where associated unilateral phrenic nerve palsy in an 11-years-old girl resulted in respiratory failure¹⁵. Cranial neuropathy is unusual (5.0%) in chronic inflammatory demyelinating polyneuropathy¹⁶. Exceptionally, facial weakness was found in 20.0% to 33.0% of the children¹⁷. In some studies, cranial nerve dysfunction has been described^{3,14,18,19}. In these children, diplopia has been a common feature^{3,18,19}. Other oculomotor symptoms comprised afferent pupillary defect and partial right sixth nerve palsy, ptosis, abduction deficit and unilateral tonic pupillary dilatation^{14,20}. Bulbar disorders resulted in dysphagia, and mastication, hypophonia and lingual fasciculations^{19,21}. Generalized facial weakness and dysarthria have also been shown^{18,19,22}. Sensorineural deafness was defined in one patient²².

Here, we reported a case of atypical CIDP with multiple cranial nerves palsy. Child presented with progressive proximal and distal weakness, along with right sided complete ophthalmoplagia, bilateral VI nerve and right sided VII nerve palsy. To our knowledge, this is the first case report of Childhood CIDP associated with multiple cranial nerve palsy in Bangladesh. Albumino cytological dissociation on CSF study and neurophysiologic findings

included conduction block, temporal dispersion, and abnormal late responses was confirmed the diagnosis of chronic inflammatory demyelinating polyneuropathy. In this case, the remarkable response to immunomodulatory therapy was convincing evidence for an acquired demyelinating neuropathy.

Conclusion

Diagnosis and management of pediatric chronic inflammatory demyelinating polyneuropathy is challenging due to the rarity of this condition and become more difficult with atypical presentation. We highlight the importance of clinical suspicion of pediatric CIPD as it is potentially treatable. Early recognition and prompt treatment can prevent long term morbidity.

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