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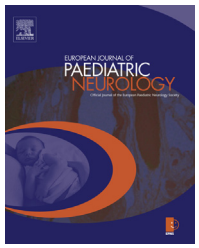

Original Article

Acute transverse myelitis in childhood: A single centre experience from North India

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ABSTRACT

Background: Acute transvers myelitis (ATM) is a rare and disabling condition in childhood. There are only few reports of clinical profile, prognosis and predictors of ATM from developing countries.

Objective: To study the clinical profile of children with ATM and predictors of its outcome.

Method: Retrospective analysis of children <12 years of age diagnosed with ATM over a period of 6 years from a tertiary care institute.

Results: Thirty six children (21 boys, median age-7.5 years) were diagnosed with ATM. Weakness was symmetrical at onset in 27 (75%) children with progression over a median of 2 days (IQR 1–5 days). Severe weakness at onset with lower limb power ≤1/5 on MRC scale was present in 27 (75%), a sensory level in 25(69.4%) and bladder dysfunction in 31(86.1%) children. MRI showed longitudinal extensive myelitis (LETM) in 27 (75%) children and the thoracic cord was most commonly affected [18 (50%)]. On a median follow up of 35 months (range IQR 11–57 months); 15 (41.7%) were non ambulatory or required assistance to walk. Severe weakness at onset with power ≤1 on MRC scale, spinal shock, respiratory muscle weakness, mechanical ventilation, greater mean time to diagnosis and treatment was associated with bad outcome. ATM was a monophasic illness in all, except in 3 children; all with neuromyelitis optica spectrum disorder. Progression to multiple sclerosis was not seen in any child in our cohort.

Conclusion: In this series of childhood ATM from North India, the disease was severe, monophasic and involved long segments (≥3) of cord in majority. Nearly half the children remain dependent on follow up. Delayed diagnosis and delayed initiation of steroid therapy was associated with poor outcome.

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1. Introduction

"Acute transverse myelitis" (ATM) describes a group of inflammatory disorders characterized by acute or sub acute onset of motor, sensory, and autonomic spinal cord dysfunction.[1]The incidence of ATM has been estimated to be between 1.7 and 2 per million children per year.[2] ATM can occur as an idiopathic monophasic illness or can herald an underlying infectious disease, or can represent the sentinel event of a chronic autoimmune disorder such as multiple sclerosis (MS) or neuromyelitis optica (NMO).[3–5] Up to 20%–50% of children may have residual motor disabilities and sphincter disturbances on long term follow up.[2,3,6,7] The American Academy of Neurology (AAN) published evidence-based ATM management guidelines focusing on two clinical settings: acute complete transverse myelitis (ACTM) and acute partial transverse myelitis (APTM).[8] This distinction is useful for consideration of underlying etiologies, prognosis, and relapse risk. These guidelines are primarily aimed at adults and similar guidelines for childhood transverse myelitis are lacking.[8]

Multiple case reports, retrospective cases series of childhood ATM have been reported from developed countries.[3,5–7,9,10] However; there is paucity of information about clinical features, etiology and outcome of ATM in children from developing countries.[11–13] Due to the different infectious triggers and variation in immune responses the profile of childhood transverse myelitis may be different in developing countries. Keeping this in mind we report our experience in childhood ATM describing clinical profile, radiological features, long term outcome and factors affecting outcome of ATM over a 6 year period.

2. Methods

This study was a retrospective review of children diagnosed with ATM between January 2008 and August 2014 at a single tertiary care children hospital of North India. The hospital is a referral, research and teaching children hospital catering to a population of more than 85 million in North India. As a policy only children less than 12 years are admitted in our hospital. All children presenting with ATM are admitted to the pediatric emergency services. They are then shifted and managed by the pediatric neurology service unless the severity of the illness calls for intensive care unit admission. After the acute phase these children are followed up in the pediatric neurology outdoor follow up clinic. The data of the clinical features, investigations, and follow up is maintained separately for each child in the clinic. For the purpose of this study, we reviewed the follow up clinic records and children diagnosed with any acquired central nervous system (CNS) demyelinating condition were shortlisted. Those with an evidence of myelitis were reviewed in detail. These included children with concomitant optic neuritis and/or encephalopathy. The review included a retrieval of hospital record file (maintained during acute hospitalization phase), and follow up clinic files. Additionally all parents and/or caregivers were contacted for a one-time follow up assessment to ascertain residual disabilities.

The diagnosis of ATM was based on the diagnostic criteria provided by transverse myelitis consortium working group (TMCWG). These include (1) sensory, motor, or autonomic dysfunction attributable to the spinal cord; (2) bilateral signs or symptoms but not necessarily symmetric; (3) clearly defined sensory level, (4) progression to nadir less than 21 days following the onset of symptoms; and (5) Exclusion of extra-axial compressive etiology by neuroimaging (MRI or myelography; CT of spine not adequate), brain abnormalities suggestive of MS.[1] Cases were defined as definite ATM if they also had presence of TMCWG inflammation criteria, which included one of (1) Cerebrospinal fluid (CSF) pleocytosis > 10 leukocytes/mm^3; (2) elevated immunoglobulin G index or presence of intrathecaloligoclonal bands; or (3) MRI spinal gadolinium enhancement. Cases were defined as possible ATM if they had absence of TMCWG inflammation criteria or if a lumbar puncture or spinal gadolinium MRI procedure was not carried out.[1,7] For this study we used a pediatric adaptation of these criteria. We omitted the criteria "*presence of a clearly defined sensory level*" because demonstration of a true spinal segment is often difficult in children.[7,11] Acute partial transverse myelitis (APTM) and acute complete transverse myelitis (ACTM) were classified based on recommended definitions.[8] Longitudinally extensive transverse myelitis (LETM) was defined as involvement of 3 or more segments of cord on MRI. The diagnosis of Neuromyelitis Optica (NMO), Neuromyelitis Optica Spectrum Disorder (NMOSD) and Multiple Sclerosis was based on criteria provided by Wingerchuk 2006 and International Pediatric Multiple Sclerosis Study Group (IPMSSG) 2010.[14,15] Spinal shock was defined as depressed spinal cord reflexes caudal to spinal cord injury.[16]

Base line demographic information, antecedent infections, vaccination, clinical features, maximum disability, duration of symptom progression, CSF findings, serological investigations, radiological features, and treatment received were systematically recorded from the files.

All children with ATM had undergone a MRI on a 1.5 T scanner. The site of lesions, distribution, extent of lesions, number of segments involved and presence of gadolinium contrast enhancement were recorded. MRI brain was reviewed for McDonald 2010 criteria for dissemination in space, as recently reviewed for use in childhood demyelination.[15,17] Time to diagnosis was defined as interval between onset of first symptom and establishment of a clinical diagnosis of ATM after presentation to hospital, and time to treatment was defined as time interval from onset of first symptom to initiation of steroid therapy. All children were treated with methylprednisolone at 30 mg/kg/day for 5 days followed by oral steroids for 4–6 weeks.

At last follow-up, diagnosis was considered as monophasic ATM (no clinical relapse or no appearance of new radiologic lesions) or relapsing ATM (MS, NMO or NMOSD as defined by IPMSSG).[15] The final outcome was categorized in 2 groups: good outcome and bad outcome. The bad outcome group consist of children who were non-ambulatory or ambulatory

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