Case Report

Short Term Outcome of Writer's Cramp in Children - A Case Series

Dipa Saha[1], Narayan Chandra Saha[2], Chinmoy Kumar Saha[3], Jobaida Parvin[4], Banita Mistry[5], Sufia Khatun Sumi[6], Husnea Ara Khan[7]

Abstract:

Background: Focal task-specific dystonia of the hand is unusual in childhood. Childhood dystonia can result in lifelong disability and thus represents a significant diagnostic and rehabilitation challenge. A detailed clinical evaluation and comprehensive work-up to rule out treatable causes of focal dystonia are mandatory for establishing a therapeutic strategy. We reported 4 cases of writer's cramp with earliest age of onset. Our aim was to focus on early detection, proper evaluation and management of writer's cramp to ensure better quality of life of an affected child.

Key words: Writer's cramp, Childhood dystonia, Focal Dystonia

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1. Dr. Dipa Saha
   Associate Professor, Pediatric Neurology
   Addin Women's Med College Hospital (AWMCH).
2. Prof. Narayan Chandra Saha
   Professor and Head Department of Pediatric Neurology
   National Institute of Neurosciences and Hospital
3. Dr. Chinmoy Kumar Saha
   Associate Professor and Head, Department of Medicine
   Comilla Medical College
4. Dr. Jobaida Parvin
   Assistant Professor, Pediatric Neurology
   National Inst. of Neurosciences & Hospital (NINS)
5. Dr. Banita Mistry
   Assistant Professor, Pediatric Neurology
   National Inst. of Neurosciences & Hospital (NINS)
6. Dr. Sufia Khatun Sumi
   Consultant, Pediatric Neurology
   National Inst. of Neurosciences & Hospital (NINS)
7. Dr. Husnea Ara Khan
   Assistant Professor, Pediatric Neurology
   National Inst. of Neurosciences & Hospital (NINS)

Address of correspondence

Dr. Dipa Saha
Associate Professor, Pediatric Neurology
Addin Women's Med College Hospital (AWMCH)
E-mail:dipasaha182@gmail.com,
Cell: 01717337891

Introduction:

Writer's cramp is an uncommon form of focal dystonia limited to small muscles of hand, is of ten action specific[1]. Symptoms may vary with unusual positioning of the fingers, wrist or elbow. It is often associated with muscle spasms and cramping while writing, with or without involuntary movement and pain. It may often be progressive and hamper other daily activities. However, a neurological examination including tendon reflexes remains normal[2]. Most of the cases have no identifiable cause. The list of possible genetic and non-genetic causes of childhood focal dystonia is extensive.

Writer's cramp causes substantial functional disability and is frequently delayed or misdiagnosed. Diagnostic work up of childhood dystonia is often challenging for clinicians. Early recognition, proper evaluation and management may improve the quality of life and better schooling of the affected child. For that reason, we reported 4 cases of childhood writer's cramp.

Methodology:

This prospectively designed cohort study was conducted among 4 children attending the outpatient department (OPD) of pediatric neurology, National Institute of Neurosciences and Hospital (NINS), Dhaka, during the period of June 2017-December 2019. Written informed consent were taken from the parents of respective child prior to enrollment, and, all parents consented to share video and pictures except 1 who agreed to share clinical information and lab reports, only. Every patient was evaluated through history, physical examination and investigation. Ophthalmological evaluation, Neuroimaging, urinary copper, serum ceruloplasmin and metabolic screening

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was done to find out underlying pathology. Genetic testing was done in affordable cases. All patients were treated with anti-dystonic drugs and their outcome was assessed after 6 months.

Results:

No gender differences existed among these 4 patients (mean age 9.66 ± 1.03years). The mean age of onset was 7.75 ± 1.78 (6-10.5) years among all 4 children. Notably, no difference existed between the younger and older age groups of children in terms of location of WC: while flexor dystonia (Rt hand), Dystonic postures of hand & forearm, and, Extensor dystonia (rt hand) among the slightly higher aged children, Flexor dystonia (Rt hand) were among the slightly lowed aged ones. None of them had positive KF ring **(Table 1)**. Mean Urinary Copper (46.83 ± 23.24) µgm/l and mean S. ceruloplasmin was 31.24 ± 5.76mg/dl those were normal for all cases, Metabolic screening were negative and Neuroimaging findings were also normal. No pathogenic or likely pathogenic variants were found on genetic study which was done among 2 cases. Another 2 cases were unable to do genetic study **(Table 2)**. On the clinical outcome among all 3 of 4 cases receiving anti-dystonic drugs at 6 months demonstrated improvement (case-1, 3, 4) but 1 case remained static (case 2). **Table-3**.

Table-I: Clinical profile among the cases

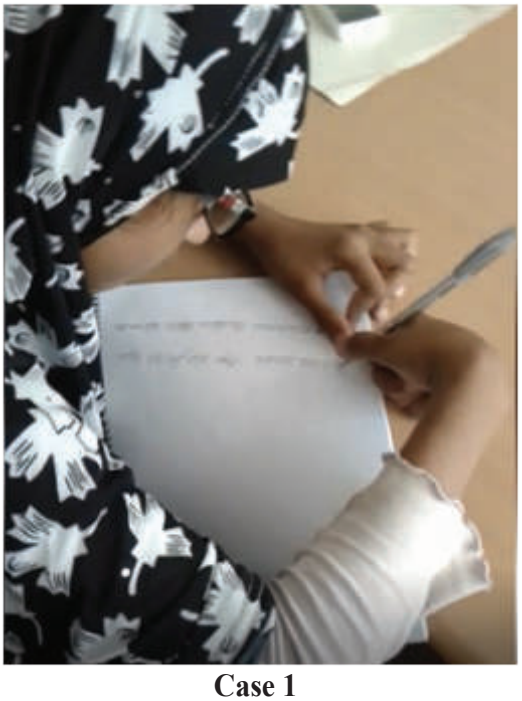
SN	Age	Sex	Age of onset	Clinical features	Ophthalmological evaluation
Case1	10 years	Female	6 years	Flexor dystonia (Rt hand)	Normal
Case2	10 years	Female	9 years	The dystonic postures of the hand and forearm, with index finger and thumb extension, wrist flexion and excessive pressure used to hold the pen	Normal
Case3	11 years	Male	10.5 years	Extensor dystonia (rt hand)	Normal
case4	9 years	Female	8 years	Flexor dystonia	Normal

Table-II: Investigation and outcome of the study cases

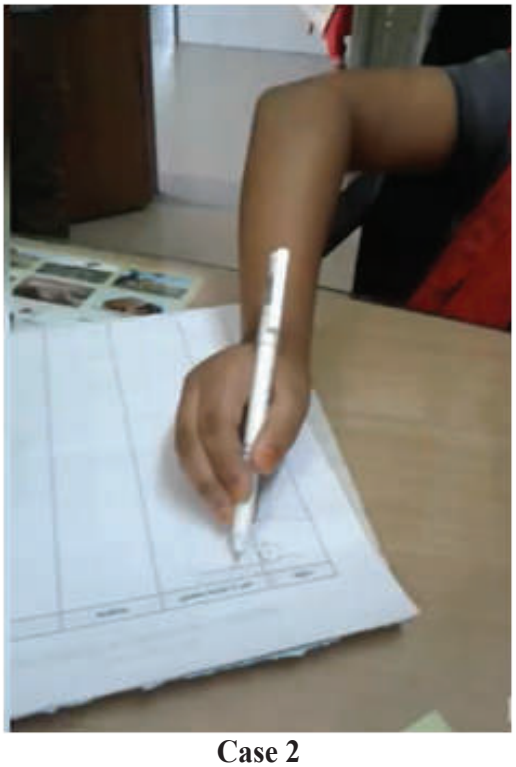
SN	Urinary Copper <100 ugm/l	Serum cerulopl asmin (20-60mg/dl)	MRI of brain	Metabolic screening (ABG, S. lactate, Blood NH_3 Urinary ketone	Genetic analysis
Case 1	56±3.9ugm/l	32.2mg/dl	Normal	Normal	no pathogenic or likely pathogenic variants
Case 2	23.6±3.1ug m/l	27.81m g/dl	Mild chronic is chemic changes in both parietal region (not clinically correlate)	Normal	Not done
Case 3	89±8ugm/l	37mg/dl	normal	Normal	Not done
Case 4	37±2.1ugm/l	25.9mg/dl	normal	Normal	no pathogenic or likely pathogenic variants

Table-III: Clinical outcome among the cases at 6 months (n=4)

SN	Treatment	Outcome
Case 1	Antidystonic drugs	Improving
Case 2	Antidystonic drugs	Static
Case 3	Antidystonic drugs	Improving
Case 4	Anti-dystonic drugs	Improving



Case 1



Case 2

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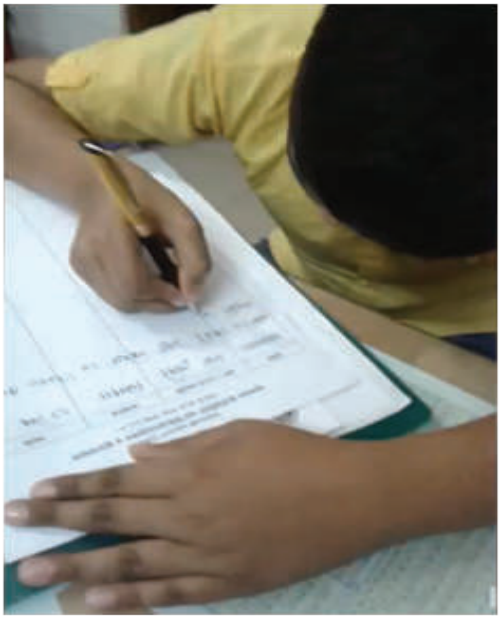
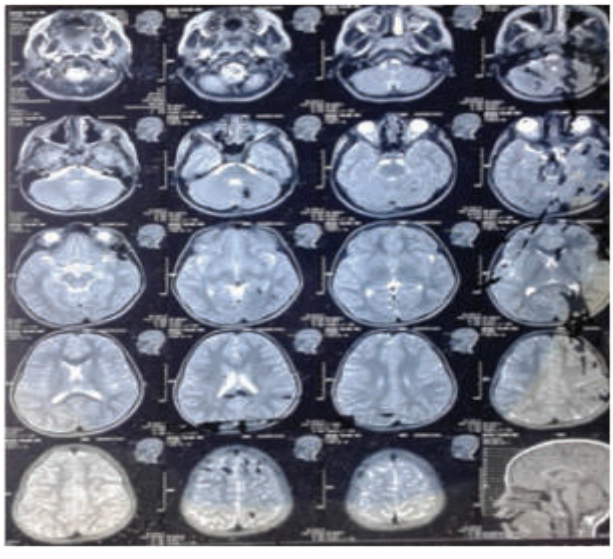
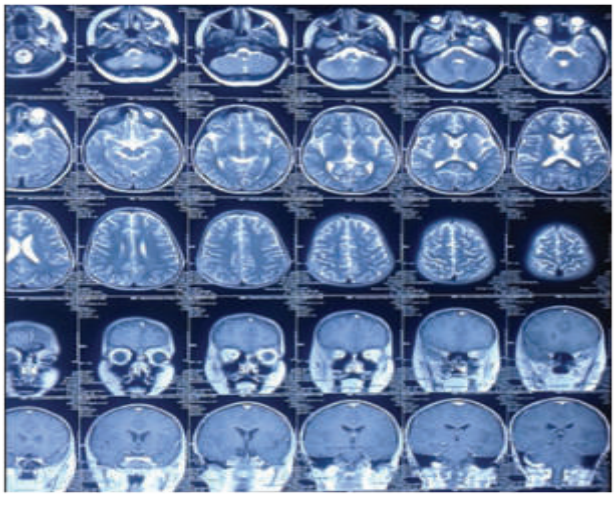


Case 3



Case1



Case 2

Normal neuroimaging among the cases

Discussion:

Writer's cramp reported mostly in adult population. The mean age of onset is 38 years and manifests as an abnormally tight grip while writing and remissions are uncommon[3].Writer's cramp in adult differs from that of pediatric population in many aspects: more often secondary than primary, mixed motor disorders (e.g., dystonia associated with spasticity), the course of dystonia may be influenced by ongoing brain maturation and by the remarkable plasticity of young brain, drug tolerability and effectiveness can be different in children[4].

Some reports of childhood writer's cramp were scarcely seen but those were of secondary origin. As one case report showed that age of onset was 8 years but underlying etiology was inherited disorder[5]. But our reported cases were of earlier onset, one presented at her age of 6 years and another one at 9 years.

Among the secondary causes of writer's cramp myoclonus-dystonia syndrome (DYT-11)[6] [7], dopa responsive dystonia and X linked dystonia-parkinsonism (DYT-3)[3], basal ganglia arteriovenous malformations[8], basal ganglia infarction[9], neuro metabolic like L-2-hydroxyglutatric[5] Wilson's disease, Huntington's disease, pantothenate kinase-associated neurodegenration (PKAN), glutaricaciduria, GM1 gangliosidosis, and Leigh's disease are most often found. But thorough investigation including genetic analysis were not enough to identify the secondary causes in our cases. So, our cases were regarded as primary writer's cramp.

Conclusion:

Childhood dystonia can result in lifelong disability and thus represents a significant diagnostic and rehabilitation challenge. A detailed clinical evaluation and comprehensive work-up to rule out treatable causes of focal dystonia are mandatory for establishing a therapeutic strategy.

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Conflict of interests: No conflict of interest

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