J Young Pharm, 2022; 14(2) : 256-257
A multifaceted peer reviewed journal in the field of Pharmacy
www.jyoungpharm.org | www.phcog.net

Case Report

A Rare Case Report on Opsoclonus Myoclonus Syndrome Associated with a Developmental Regression

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ABSTRACT

Developmental regression can be described as the progressive loss of previously acquired skills which is relatively rare. Opsoclonus myoclonus syndrome is an uncommon autoimmune disorder in which a patient's body attacks its own nervous system. It usually affects young children. Complicated developmental regression may lead to diagnosis of Opsoclonus Myoclonus Syndrome (OMS). A 3 years and 1 month old, the female child presented the history of convulsion, tonic contraction of upper and lower limb, up rolling of eye ball, frothing from mouth and post ictal drowsiness at the age of 1.5 years. On examination her general condition was unstable and systemic examination was not normal (Temp- 102.2 F). Her hematological and hepatic tests were done. Magnetic Resonance Imaging (MRI) report of her brain conclude that few T2 and FLAIR hyper intense foci are noted in bilateral periventricular deep white matter possibility of myelin pallor likely. We show that Opsoclonus myoclonus syndrome can be associated with developmental regression. Neurologist must be aware of such presentation. Our findings highlighted the need for increased recognition of developmental regression leading OMS as an urgent and treatable condition.

Key words: Opsoclonus myoclonus syndrome (OMS), Developmental regression, Paraneoplastic, Myelin pallor, Ataxia, Cerebellar nuclei degeneration.

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DOI: 10.5530/jyp.2022.14.49

INRODUCTION

Developmental regression can be described as the progressive loss of previously acquired skills which is relatively rare. Where a patient had normal development of activities (communicative and linguistic abilities or activities for example smiling, eating, attentiveness etc) previously and progressively loses the key 'milestone'. For instance, a child who became able to walk at 13 months, and then at 3 and half year become unable to walk then unable to speak and so on. For this specific condition many other alternative terms are used such as "progressive intellectual and neurological deterioration" (PIND). There are wide range of etiological factors and idiopathic conditions which are responsible for causing regression ex., psychological/environmental factors (psychological trauma may lead to patient to behave in age-inappropriate manner ex., a toddler speak in a way of 'baby language'), chronic acute infection under the name of non-neurological disease (for example Urinary Tract Infection associated with loss of continence), neurological disorders (space-occupying lesion, autism, seizure attacks etc), other genetic neurodegenerative conditions (for ex., classical Rett syndrome). There are no particular laboratory tests for diagnosing the regression but diagnosis can be done through observing behavior changes during examination and other radiology tests such as Computed Tomography (CT) scan and MRI.

Opsoclonus myoclonus syndrome is a uncommon autoimmune disorder in which patient's body attacks its own nervous system. It usually affects the young children. The estimate incidence rate is one in five million individuals all over the world. Furthermore most of the people with OMS have neuroplastic, neuroblastoma, small benign, viral infections, metabolic or unknown etiologies. Symptoms mainly involved rapid and repeated eye movement (Opsoclonus) and Muscle jerks (Myoclonus) hence it is also far-famed as 'dancing eye' and 'dancing feet' syndrome. Other symptoms involved are ataxia i.e. loss of balance, inability to stand, walk, sit, and crawl. Apart from this patients developed hand tremors, sleep problems, behavior problems and vomiting. It can be diagnosed through medical history and examine nervous system through radiological tests ex., MRI, CT Scan and Cerebrospinal fluid (CSF) testing for any kind of inflammation in brain or spinal cord. Most of the treatments consist of Immunosuppressive drugs(Corticosteroids, Adrenocorticotropic hormone, Intravenous immune globulin – IVIG, Cyclophosphamide, Rituximab).[1]

In some cases, complicated developmental regression may lead to diagnosis of Opsoclonus Myoclonus Syndrome (OMS). This condition is rarely seen in younger children of about 1-4 years. In this type of diagnosis patient experienced frequent myoclonic jerk which disappear during sleep and also feeling of frustration.[2]

CASE REPORT

A 3 years and 1 month old, female child presented in multispecialty hospital (date of admission- 03/08/2021) with the history of convulsion, tonic contraction of upper and lower limb, uprolling of eye ball, frothing from mouth and post ictal drowsiness at the age of 1.5 years. She had one more seizure episode after one month. For the above complaints patients went to multiple Out Patient Department (OPD) centers. Patient were prescribed with syrup VALPARIN (Sodium Valproate) 2.5 ml twice a day and tablet SERENACE (Haloperidol) 0.25mg 1/4th Once a day in February 2020. Furthermore, patient visited multiple OPD centers for the treatment but was not cured. She has one and half year old brother. Both pregnancies and birth were without complications. Patient weight was 3 kg at the time of birth. No one in the family has similar history.

She was presented with the complaints of not able to walk, stand, crawl, speak, abnormal movements (both the limbs), constant gradually

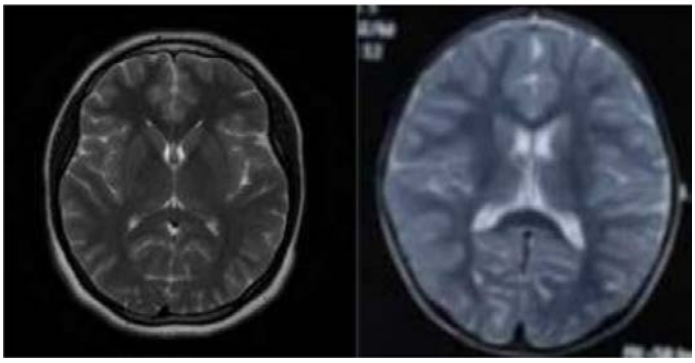

Figure 1: Normal vs abnormal patient's MRI Scan.

progressive jerky movements which disappear while asleep, uprolling eye ball, abnormal blinking. Her mother noted that she was a happy child who used to do eye contact, walking, standing, waved 'bye-bye' and played until 1.5 year from this time her behavior became progressively deteriorate. Her walking became unstable and eye contact almost disappeared. She did not played with toys or other objects and engaged in confused and repetitive behavior. She had involuntary random, bilateral, conjugate eye movements consist with Opsoclonus, myoclonus (Proximal < Distal limbs) and continous tremors. She was not able to sit upright due to Ataxia.

On examination her general condition was unstable and systemic examination was not normal (Temp- 102.2 F). Her hematological and hepatic tests were done and the reports were almost normal. Radiological tests like MRI, CT SCAN, Ultrasonogram (USG) were done and diagnosis was made. MRI report of her brain conclude that Few T2 and FLAIR hyper intense foci are noted in bilateral periventricular deep white matter possibility of myelin pallor likely. Figure 1 CT-Thorax (plain) showed that area of consolidation with surrounding ground glass haziness with air bronchograms are noted involving depended parts of bilateral lungs filled. Contrast-Enhanced Computed Tomography (CECT) scan of abdomen and pelvis showed liver measured 9.2 cm appear increased in size suggestive of hepatomegaly, spleen measured 7 cm appear in size suggestive of splenomegaly.

Patient was admitted for 12 days for suspected manifestation. She was given with following symptomatic treatment: Inj. 0.9 DNS + 5cc KCL (41 ml/hour) IV, Inj. IVIG (10 gm/100 ml) continuously 6 to 8 hr. Inj. ACTH (60 IU/ml) IM 12 hr, Tab. Clobazam ½ tablet (2.5 mgin 5ml water) PO 24 hr, Tab. Clonazepam ½ tablet (0.25 mg in 5 ml water) PO 12 hr, and Tablet. Folic acid (5 mg) PO thrice a week. Syp. Levetiracetam (3 ml) PO 12 hr, Syp. MVBC (5 ml) PO 12 hr, Syp. Augmentin (4 ml) PO 8 hr. 0.9 DNS + 5 cc KCL was given for the electrolytes balance. Inj IVIG was prescribed as immunosuppressive. ACTH was given for convulsive movements. Tablet Clobazam was given for sedation. Tablet Clonazepam and syrup Levetiracetam were given for seizure attacks. Tablet Folic acid and tablet Multivitamin + B complex (MVBC) were prescribed as multivitamins. Syrup Augmentin was prescribed for bacterial infection.

DISCCUSION

This case study depicts how continues developmental of regression leads to Opsoclonus Myoclus Syndrom (OMS). This autoimmune condition is rare, which is characterized by cerebellar nuclei degeneration. It occurs frequently as a paraneoplastic syndrome. Cerebellar neurons are poisoned by chemicals produced by neoplastic cells. Opsoclonus, Myoclonus, and Ataxia are common clinical symptoms of this neurological disorder before the fundamental aetiology is discovered. Opsoclonus is characterized by uncontrollable vertical and horizontal eye movements. Myoclonus is characterized by sudden muscle jerks. This condition includes dyspraxia (developmental co-ordination deficit), dysphagia (swallowing difficulty), and dysarthria (slurred speech), as well as hypotonia, lethargy, and malaise in the cerebellum. Half ofthe all OMS cases occur in children with a neuroblastoma, with onset often before 4 years of age. Children with OMS typically have chronic, disabling cognitive and neurological consequences. OMS can be clinically diagnosed with a physical examination. Patients with OMS should have a full physical examination to rule out malignancy and infections. Additional factors in determining the projection are the age of onset, stage and type of the infection, degree of neurological involvement, stage of tumor eradication, the effectiveness of treatment. Relevant counseling to the patient / patient's family in such cases found to be beneficial.[3,4]

CONCLUSION

In this case report we accentuate a 3 years and 1 month old female patient suffering from acute cerebellar ataxia which is nearly universal misdiagnosis, the index for OMS should be increased in the children specific less than 2-4 years with tread ataxia, anger and impatience, even before the occurrence of OMS. The MRI, CT Thorax, USG, CECT tests were done and may provide an influential clue for OMS and neuroblastoma. In this case MRI findings indicates significant abnormalities. Our findings highlighted the need for increased recognition of developmental regression leading OMS as an urgent and treatable condition.

ACKNOWLEDGEMENT

The authors are sincerely thankful to all the members of Department of Pharmacy, Sumandeep Vidyapeeth (Deemed University) (SVDU) for their immense support and guidance.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ABBREVIATIONS

MRI: Magnetic Resonance Imagine; **OMS:** Opsoclonus Myoclus Syndrome; **PIND:** progressive intellectual andneurological deterioration; **CSF:** Cerebrospinal fluid; **USG:** Ultrasonogram; **OPD:** Out Patient Department; **CECT:** Computed Tomography.

REFERENCES

1. Jiménez Legido M, Cantarín Extremera V, Fournier del Castillo MdlC, Melero Llorente J, González Gutiérrez-Solana L. Síndrome opsoclonus mioclonus. Experiencia en los últimos 12 años en un hospital terciario. Anales de Pediatría. 2020;93(5):339-42. doi: 10.1016/j.anpedi.2019.12.007.
2. Garg RK, Rizvi I, Malhotra HS, Kumar N. Opsoclonus myoclonus syndrome in children: Paraneoplastic versus parainfectious. Neurol India. 2018;66(5):1295-8. doi: 10.4103/0028-3886.241387, PMID 30232990.
3. Taib BG, Kinshuck AJ, Milburn-McNulty P, Fratalia L, Forsyth L, Husband D, *et al.* Opsoclonus-myoclonus syndrome associated with a nasopharyngeal tumor in an adult: a case report. J Med Case Rep. 2015;9(1):128. doi: 10.1186/s13256-015-0605-9, PMID 26033370.
4. Scarff JR, Iftikhar B, Tatugade A, Choi J, Lippmann S All from University of Louisville School of Medicine. Opsoclonus myoclonus. Innov Clin Neurosci. 2011;8(12):29-31. PMID 22247816.

Article History: Received: 31-01-2022; Revised: 18-03-2022; Accepted: 30-04-2022.
Cite this article: Suratiya N, Shah K, Soni P, Khare S. A Rare Case Report on Opsoclonus Myoclonus Syndrome Associated with a Developmental Regression. J Young Pharm. 2022;14(2):256-7.