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A CASE REPORT ON OPSOCLONUS MYOCLONUS ASSOCIATED WITH ATAXIA

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Abstract

Introduction: Opsoclonus-Myoclonus-Syndrome is a rare autoimmune disorder in which the body targets the nervous system, causing a variety of symptoms. Presented with triad of symptoms Opsoclonus, myoclonus, ataxia and is most often associated with tumors or infection. Most toddlers are misdiagnosed with childhood acute cerebral ataxia instead of Opsoclonus-myoclonus-ataxia syndrome.

Case Report: A 3.5 year old male presented with complained of being unable to walk, stand, crawl, or speak, as well as strange movements (both limbs), persistent gradually escalating jerky movements that vanished while he was sleeping, up rolling of the eyeball, and irregular blinking. Magnetic resonance imaging revealed that Few T2 and FLAIR hyper intense foci are noted in bilateral periventricular deep white matter possibility of myelin pallor. The treatment started with immunosuppressant and anticonvulsants.

Conclusion: We show that Opsoclonus-myoclonus associated with ataxia in children is usually misdiagnosed with childhood acute cerebral ataxia. Our findings emphasised the need of increasing awareness of OMS accompanied with ataxia as a serious and curable illness. We feel our findings are important because they give clinical experience that might aid in a better understanding of this condition.

Key Words: Opsoclonus, Myoclonus, Cerebral Ataxia, Paraneoplastic.

INTRODUCTION:

Opsoclonus-myoclonus syndrome, which affects children, is a life-threatening neuroinflammatory condition that is frequently paraneoplastic. OMS (dancing eye-dancing feet syndrome) is another name for it. It is usually seen in infants and young children, it rarely affects adults. Opsoclonus is a condition in which the eyes move in a seemingly random, involuntary, and quick manner in the horizontal, vertical, and diagonal directions. Myoclonus refers to limb jerks or lightning-like motions which can also be tremulous. Ataxia is another symptom of the disease, which tends to result in the inability to walk, sit, or

crawl, as well as the loss of previously acquired functions such as speech.OMS is assumed to be an autoimmune disorder in which the body targets the nervous system, causing a variety of symptoms.

The exact etiology of OMS is unknown, but it is frequently caused by paraneoplastic disease in kids. The theory is that a concealed or occult tumor (such as neuroblastoma or ganglioneuroblastoma) activates the immune system, and the ensuing antibodies unknowingly target healthy nervous system cells. CD20+ B-cells seen in the CSF fluid appear to play a key function. The cerebellum, which is responsible for coordinating body motions, is one of the brain areas that is impacted. OMS is thought to be initiated by infections in older children and adolescents, and it is thought to be triggered by paraneoplastic processes in adults, such as lung or breast cancer.

OMS is frequently referred to as OMAS- with 'A' presenting ataxia- because ataxia is always present in children with OMS. It's also known as Kinsbourne syndrome, after Marcel Kinsbourne, who originally described it in an infant in 1962. In reality, most toddlers are misdiagnosed with childhood acute cerebral ataxia. Symptoms such as Opsoclonus and myoclonus, which can start suddenly and progress over days to weeks, are frequently used to diagnose OMS. Other symptoms include lack of developmental milestones such as speech, high irritability, and significant sleep difficulties. [1]

Although no diagnostic test exists, a lumbar tap can be used to detect for antibodies known as oligoclonal bands (OCBs) in the cerebrospinal fluid (CSF). Additionally, lymphocyte flow cytometry can be used to look for increased CD20+ B-cell numbers in the CSF. To help identify malignancies that may have induced OMS, a CT scan, MRI scan, PET scan, or a combination of these scans can be used. It's also crucial to rule out other causes of symptoms, such as a brain tumor or leisions, which can be detected with an imaging study.

CASE REPORT:

A 3 year and 5 months old male kid presented to a multispecialty hospital (date of admission- 07/09/2021) with a history of convulsion, tonic contraction of upper and lower limb, up rolling of eye ball, frothing from mouth, and post ictal lethargy at the age of 1.5 year. After one month, he suffered another seizure. Patient visited several Out Patient Department (OPD) centers for the aforementioned problems. In February 2020, the patient was given the syrup VALPROL (Sodium Valproate) 2.5 ml twice a day and the pill HALOBID (Haloperidol) 0.25mg 1/4th once a day. Furthermore, the patient sought therapy at different OPD centers but was not cured. His younger brother is one and a half years old. Both pregnancies and deliveries went smoothly. At the moment of birth, the patient weighed 3.8 kg. There is no one else in the family with a similar medical background.

He complained of being unable to walk, stand, crawl, or speak, as well as strange movements (both limbs), persistent gradually escalating jerky movements that vanished while he was sleeping, up rolling of the eyeball, and irregular blinking. His mother described him as a cheerful kid who used to make eye contact, walk, stand, wave "bye-bye," and play until he was 1.5 years old, when his behavior began to degrade. His walking became unsteady, and he lost eye contact nearly completely. He didn't play with toys or other objects, and his behavior was confused and repetitious. Opsoclonus, myoclonus (proximal < distal limbs), and continuous tremors were among his involuntary random, bilateral, conjugate eye movements. Due to his ataxia, he was unable to sit up straight.

On examination his general condition was unstable and systemic examination was not normal (Temp-102.2 F). His hematologic and hepatic investigations were conducted, with outcomes that were nearly normal. Radiological testing such as MRI, CT SCAN, and USG were performed, and a diagnosis was made. MRI report of his brain concludes that Few T2 and FLAIR hyper intense foci are noted in bilateral periventricular deep white matter possibility of myelin pallor likely. CT-Thorax (plain) revealed a region of consolidation with surrounding ground glass haziness, as well as air bronchograms, implicating dependent sections of the bilateral lungs. 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 7cm appear in size suggestive of splenomegaly.

The patient was admitted to the hospital for a period of 12 days because to a suspected manifestation. The following symptomatic therapy was prescribed for her: Inj. 0.9 DNS + 5cc KCL (41 ml/hour) IV, Inj. IVIG (10 gm/100 ml) continuously 6 to 8 hourly. Inj. ACTH (60 IU/ml) IM 12 hourly, Tab. Clobazam ½ tablet (2.5 mg in 5ml water) PO 24 hourly, Tab. Clonazepam ½ tablet (0.25 mg in 5 ml water) PO 12 hourly, Tab. Folic acid (5 mg) PO thrice a week. Syp. Levetiracetam (3 ml) PO 12 hourly, Syp. MVBC (5 ml) PO 12 hourly, Syp. Augmentin(4 ml) PO 8 hourly. For the electrolytes balance, 0.9 DNS + 5 cc KCL was administered. IVIG injections with immunosuppressive properties were prescribed. ACTH was used to treat convulsive motions. Clobazam was delivered in the form of a sedative pill. Clonazepam pills and Levetiracetam syrup were used to treat seizures. Folic acid tablets and Multivitamin + B complex (MVBC) tablets were prescribed as multivitamins. A bacterial infection was treated with Augmentin syrup.

DISCUSSION

This case study explains how past seizure attacks in children lead to OMS syndrome. This is a kind of autoimmune disease. Also, infrequent condition. This is signalized by degeneration of the cerebellar nuclei. As a paraneoplastic syndrome, it occurs often. Chemicals released by malignant cells harm cerebellar neurons. Before the underlying aetiology of this neurological condition is determined, typical clinical signs include opsoclonus, myoclonus, and ataxia. Uncontrollable vertical and horizontal eye movements are a symptom of opsoclonus. Myoclonus is characterised by muscular twitches that occur suddenly. Dyspraxia (developmental co-ordination loss), dysphagia (swallowing difficulties), and dysarthria (slurred speech) are all symptoms of this disorder, which also involves hypotonia, lethargy, and malaise in the cerebellum. Children with neuroblastoma account for half of all OMS cases, with onset generally occurring before the age of four. Chronic, debilitating cognitive and neurological sequelae are common in children with OMS. A physical examination can be used to diagnose OMS.A comprehensive physical examination should be performed on patients with OMS to rule out cancer and infections. The age of onset, stage and kind of infection, degree of neurological involvement, stage of tumour elimination and therapy efficiency are all variables in determining the projection. In such circumstances, relevant counselling to the patient or the patient's family has been proven to be useful. [2]

Opsoclonus myoclonus syndrome is a disorder caused by the immune system. Early therapy and proper intensification may have an influence on medium to long-term results in some patients linked with an underlying tumour (particularly in terms of cognitive development).due to all of the above, despite its low incidence. This condition should be considered in patients who develop any of its usual symptoms suddenly, notably ataxia. [3]

We also found anotherarticle that is similar to our case report, under the title of “opsoclonus myoclonus syndrome in children: Paraneoplastic versus para infectious”. They show paediatrics age group in this article. And manifest two predominant etiological factors it may either be paraneoplastic or para infectious (that is infective or post infective). In both conditions the pathogenesis is immune mediated. In this study the group of 14 children (age of 5 years or less) was involved. And in 8 cases, the common referral diagnosis was acute cerebellar ataxia. Opsoclonus myoclonus along with ataxia and irritable behaviour were the most common clinical features. [4]

CONCLUSION

In this case report, we focus on a 3 year 5 months old male patient who has Opsoclonus-myoclonus-ataxia syndrome. The index for OMS should be raised in children aged 2-4 years who have severe ataxia, anger, and impatience, even before the onset of OMS. If we look at the progression of patients through time and the treatment regimens utilised, it appears that a better knowledge of the illness, as well as the necessity of early therapy and intensification, may have influenced how we manage OMS. Our findings emphasised the need of increasing awareness of OMS accompanied with ataxia as a serious and curable illness. We

feel our findings are important because they give clinical experience that might aid in a better understanding of this condition.

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