Opsoclonus Myoclonus Ataxia Syndrome
Case Reports

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Abstract:

This paper describes four cases of Opsoclonus myoclonus syndrome (OMS), which is a rare autoimmune neurological disorder. Patients present with irritability, ataxia and chaotic eye ball movements. There are no specific features differentiating paraneoplastic OMS from non paraneoplastic OMS. Acute symptoms are responsive to immuno-modulatory treatment but long term follow up can reveal neurological (mainly cognitive) sequels.

Keywords

Opsoclonus Myoclonus Syndrome, ataxia, chaotic eye ball movements, neuroblastoma

Introduction

Opsoclonus myoclonus syndrome (OMS) is a rare neurological disorder of unknown causes which appears to be the result of an autoimmune process involving the nervous system. It is an extremely rare condition, affecting as few as 1 in 10,000,000 people per year. It affects 2 to 3% of children with neuroblastoma.(1)

OMAS consists of three main symptoms:

1. Opsoclonus (conjugate, multidirectional, chaotic eye movements),
2. Myoclonus (nonepileptic limb jerking that can also involve the head and face)
3. Trunkal ataxia, which cause gait imbalance.

In addition sleep disturbance, cognitive dysfunction, and behavioral changes are often found (2)

Age of onset is typically before 3 years of age. OMAS is generally a para-neoplastic or para-infectious entity. Neuroblastoma (NB) seen in 50% of cases. 2% and 3% of children with NB have OMAS. [3]

Patients with NB and OMAS have good survival rates. 70%–80% of these children will have long-term neurologic sequels. [4]

Associations

1. Neuroblastic tumours like, neuroblastoma, ganglioneuroblastoma, ganglioneuroma, ovarian teratoma or hepatoblastoma rarely [5]
2. Flu vaccine
3. Infection like Streptococcus EBV, Mycoplasma pneumoniae, St. Louis virus, varicella rarely malaria, hepatitis C, and HIV (6, &7)

Diagnostic Criteria

International consensus has described three of the following four diagnostic criteria

(1) Opsoclonus,
(2) Myoclonus/ataxia,
(3) Behavioural change and/or sleep disturbance, and
(4) Neuroblastoma. (8)

Treatment

In children, OMS resolves with ACTH, or corticosteroid, but, symptoms recur after withdrawal. Other options are:

(1) Clonazepam, Baclofen, Valproate, and 5- hydroxytryptophan had provided much needed symptomatic relief to these patients.
(2) ACTH, IVIG, and Rituximab as per National Organization for Rare Disorder
(3) Steroid - Pulse methyl prednisolone and oral prednisolone.
(4) Gabapentin -in controlling eye movements in resistant Opsoclonus

Case Reports

We present a retrospective study of 4 children attending our OPD (2015-18) in which diagnosis of OMS was made on set criteria. (8). The objective of this study was to describe the clinical

profile and outcome of this disorder. The medical records of patients were retrieved and reviewed. Details of clinical symptoms, investigations, and treatment were collected.

Case 1

Presentation: Sandhaya, 1.6 year old female, weight 9.3 kg presented with excessive crying, irritability, ataxia and chaotic eye ball movement.

Investigations:

Abdominal USG was normal.

CT showed Thoraco lumbar Neuroblastoma at D2 level.

Urine - VMA absent.

Opsoclonus Myoclonus Ataxia Syndrome (Tumour) NB

Treatment: Surgery, Methyl Prednisolone x 5 followed by Prednisolone, Rivotril bid for 1month.

Case 2

Presentation: Sameer, 1 year-old male, weight 8.6 kg presented with excessive crying, irritability, trunkal ataxia, abnormal eye ball movement.

Investigations:

CT - Chest, Pelvis and abdomen normal

MRI brain normal

CSF- Herpes Simplex PCR normal

Urine- VMA absent

Opsoclonus Myoclonus Ataxia Syndrome (Non tumour)

Treatment: Methyl Prednisolone x 5 followed by Prednisolone, Rivotril bid x 1m

Case 3

Presentation: Leena, 6 year old female, weight 16 kg came with fever, instability of gait, jerky movements of limbs, head and trunk with abnormal eye ball movement.

Investigations:

CT - Chest, Pelvis and abdomen survey normal

MRI brain normal

CSF- Herpes Simplex PCR normal

Urine- VMA absent

Opsoclonus Myoclonus Ataxia Syndrome (Non Tumour)

Treatment: Methyl Prednisolone x 5 followed by Prednisolone, Rivotril

Case4

Presentation: Purushottam, 13months old, male, weight 9 kg presented with irritability, jerky movements of limb, head, neck, abnormal chaotic eye ball movement.

Investigations:

CT - Chest, Pelvis and abdomen normal

MRI brain normal

CSF- Herpes Simplex PCR normal

Urine-VMA absent

Opsoclonus Myoclonus Ataxia Syndrome (Non tumour)

Treatment: Methyl Prednisolone x 5 followed by Prednisolone

Discussion

Four patients were studied. Male-female ratio was 2:2. The minimum age of presentation ranges from 12 months to 6 years. A follow-up period of 1 to 3 years was taken. Abnormal eye movements and non epileptic limb jerking invoking head and trunk was the preponderant inaugural feature. In one patient neuroblastoma was identified. In rest 3 no cause be detected and was kept in idiopathic group. All patients received immuno-modulatory treatment. Complete recovery of OMS symptoms was obtained in all children. Comparing the patient with NB and the idiopathic group, there were no differences in age of onset, sex ratio, main presenting symptom and responsiveness to treatment. However, developmental delay issues were noted in 3, 2 of idiopathic and 1 of NB group. Neurological sequels were equally found in patient with neuroblastoma.

Conclusion

In a patient presenting with irritability, ataxia always give time to observe eye ball movements, (This is a basic clinical tip for practitioners). Smallest Clinical Clue gives you a big diagnosis.

High clinical alert, not to miss Opsoclonus. No specific features differentiating paraneoplastic OMS from non paraneoplastic OMS. Acute symptoms are responsive to immuno-modulatory treatment but long term follow up can reveal neurological (mainly cognitive) sequels.

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