Case Report

Indian Journal of Obstetrics and Gynecology
Volume 7 Number 4 (Part - I), October – December 2019
DOI: http://dx.doi.org/10.21088/ijog.2321.1636.7419.7

Opsoclonus Myoclonus Ataxia Syndrome: A Tounge Twister

Amit Rajwade[1], Divya R[2], Seeru Garg[3], Anita Soni[4]

How to cite this article:
Amit Rajwade, Divya R, Seeru Garg, *et al*. Opsoclonus Myoclonus Ataxia Syndrome: A Tounge Twister. Indian J Obstet Gynecol. 2019;7(4 Part-1):547–549.

[1]Junior Resident, [2]Secondary DNB Student, [3]Junior Consultant, [4]Consultant, Department of Obstetrics and Gynaecology, Dr LH Hiranandani Hospital, Powai, Mumbai, Maharashtra 400076, India.

Corresponding Author: Anita Soni, Consultant, Department of Obstetrics and Gynaecology, Dr LH Hiranandani Hospital, Powai, Mumbai, Maharashtra 400076, India.

E-mail: anita.soni8@hotmail.com

Received on 01.02.2019; **Accepted on** 15.10.2019

Abstract:

Paraneoplastic syndrome occurs as a consequence of tumor in the body, is not due to local presence of tumor but is due to humoral response released by tumor cells or by an immune response against the tumor. It commonly arises from tumor of lung, breast, ovaries and lymphatic system. A 25 year married female with no gynaecological symptoms came with complaints of involuntary jerky movements of both eyes with weakness and numbness in bilateral upper and lower limbs and instability in gait since 1 week. She had fever 3 months back. Initially considered post viral demyelinating disease. Symptoms worsened and in suspicion of Opsoclonus- Myoclonus- Ataxia syndrome, imaging studies and tumor markers done to rule out any tumors in the body. USG showed left adnexal mass which was confirmed on PET scan to have hot spots. Left salpingo-oophorectomy done which on HPE showed benign ovarian teratoma. Post operatively she improved symptomatically.

Keywords: Opsoclonus myoclonus; Ataxia; Nulligravida; Antiepileptics; Steroids,

Introduction

Opsoclonus myoclonus ataxia syndrome is a neurological paraneoplastic syndrome, i.e. cancer related syndromes that can affect any part of the body which are mediated by the immune response triggered by neuronal components from the tumor antigens. These antigens may be intracellular antigens which are usually cellular immune response related or cell surface antigens which are due to humoral immune response. These syndromes usually presents as symptoms before tumor detection with rapid deterioration of neurological deficits and prompt tumor control improves the neurological outcome. Diagnosis is by clinical, radiological and/or CSF findings of presence or absence of the antibodies.[1]

Opsoclonus is involuntary intersaccadic eye movements in all directions. Opsoclonus myoclonus ataxia syndrome is usually associated with lung and breast cancer in adults, neuroblastoma in children and ovarian teratoma in adolescents. It is due to the disinhibition of the fastigius nucleus of the cerebellum and anti Ri antibodies may be present occasionally. It may lead to muscle rigidity, laryngeal spasms, autonomic dysfunction, encephalopathy, coma and death. Residual psychomotor retardation, sleep and behavioral problems may persist. Treatment is by removal of the tumor, steroids, ACTH, intravenous immunoglobulin, plasma exchange and anti CD 20 antibody- rituximab.[2]

Case Report

A 25-year nulligravida married for 8 months came to neurology outpatient department with complaints of involuntary bilateral eye movements, imbalance in gait, persistent vomiting and bilateral upper and lower limb weakness for 2 days. She had a history of travel one week back fever 3 months back and no history of recent vaccination. No significant neurological or other medical or surgical past history in her and her family. She had a normal menstrual history.

At admission to hospital she was vitally stable with imbalance in gait, opsoclonus, significant truncal and gait ataxia with other motor, sensory systems, reflexes and meningeal signs being normal.

Initially considered a postviral demyelinating disease (with her past history of fever) she was evaluated and treated for the same. Her CBC, RFT, LFT, coagulation profile, CSF studies were found to be within normal limits. MRI brain with contrast revealed no abnormality. With worsening symptoms, development of myoclonic jerks, a suspicion of paraneoplastic syndromes was made by our neurologist and we started looking out for a visible tumor or occult malignancy. She was started on steroids, antiepileptics, and intravenous immunoglobulin in suspicion of the same.

During the search for the tumor, ultrasound of the pelvis showed a left ovarian solid mass probably dermoid of 7.5 × 5.8 × 5.7 cm. This is when a gynecology reference was made and we got ovarian tumor markers done –all of which were found to be normal.

A PET–CT was done to confirm if this tumor possessed the neural components responsible for the neurological symptoms. Malignant transformations and neuronal components take up increased FDG on a PET scan. The ovarian mass showed an increased FDG uptake. With the possibility of malignancy as the cause of increased uptake being ruled out by the normal levels of the tumor markers, the etiology of the neurological symptoms was concluded to be the neuronal component from the ovarian mass.

Laparoscopic left salpingo-oophorectomy was done. Histopathology showed it to be a benign teratoma. Her postoperative period was uneventful. She was discharged on day 9 with oral steroids and antiepileptic. At one week follow up, her symptoms were better and at one month follow up she was completely asymptomatic. Her steroids were tapered and antiepileptics were continued.

Discussion

Opsoclonus Myoclonus Syndrome (OMS), also known as Opsoclonus-Myoclonus-Ataxia (OMA), is a rare neurological disorder of unknown cause 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 and has been reported to occur with celiac disease and diseases of neurologic and autonomic dysfunction.

Thaís Armangué *et al*. did a retrospective cohort study and laboratory investigations of 114 adult patients with OMS at a center for autoimmune neurological disorders done between January 2013 and September 2015. They found that of the 114 patients (62 (54%) female; median age, 45 years; interquartile range, 32–60 years), 45 (39%) had P-OMS and 69 (61%) had I-OMS. In patients with P-OMS, the associated tumors included lung cancer (n=19), breast cancer (n=10), other cancers (n=5), and ovarian teratoma (n=8); 3 additional patients without detectable cancer were considered to have P-OMS because they had positive results for onconeuronal antibodies.[1]

Jitendra Sahu and Kameshwar Prasad also did a study about OMS in the years 2011 where they found out that the opsoclonus–myoclonus syndrome is a rare and distinct neurological disorder characterized by rapid multidirectional conjugate eye movements (opsoclonus), myoclonus and ataxia, along with behavioral changes in adults and irritability in children. Sometimes it is due to self-limiting presumed para-infectious brainstem encephalitis but it may also represent a non-metastatic manifestation of neuroblastoma in children and small cell carcinoma of the lung in adults.[2]

Mary Kurian and colleagues in their study of a series of 100 patients observed that opsoclonus myoclonus ataxia was found in mostly young females, and in approximately half of them, an ovarian teratoma is found.[3]

J Scholz, P Vieregge, and C Ruff had, in the year 1994, documented a case of OMS in a 45-year-old female with metastatic ovarian carcinoma. She developed OMS as a paraneoblastic syndrome. She was treated with steroids and chemotherapy following which she had complete resolution of symptoms.[4]

In conclusion, OMS is a paraneoblastic syndrome associated with ovarian tumor, mostly teratoma. It is treatable syndrome and the patient should be started on steroids, antiepileptics, and intravenous immunoglobulin and the evidence of ovarian any other tumor (breast/lung commonly) should be searched for. Early detection, prompt treatment and proper follow up is the key for treatment of OMS.

Compliance with Ethical Standards

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent was obtained from all individual participants included in the study.

Conflict of interest: There is noconfict of interest between the authors.

References

1. Armangué T, Sabater L, Torres-Vega E *et al.* Clinical and Immunological Features of Opsoclonus-Myoclonus Syndrome in the Era of Neuronal Cell Surface Antibodies. JAMA Neurol. 2016 Apr;73(4):417-24. doi:10.1001/jamaneurol.2015.4607
2. Sahu JK, Prasad K. The opsoclonus–myoclonus syndrome. Practical Neurology. 2011;11:160–66.
3. Kurian M, Patrice H. Lalive, Josep O. Dalmau, *et al.* Opsoclonus-Myoclonus Syndrome in Anti–N-Methyl-D-Aspartate Receptor Encephalitis Arch Neurol. 2010 Jan;67(1):118-21.
4. Scholz J, Vieregge P, and Ruff C. Paraneoplastic opsoclonus-myoclonus syndrome in metastatic ovarian carcinoma. J Neurol Neurosurg Psychiatry. 1994 Jun;57(6):763-4