

# Opsoclonus-Myoclonus Syndrome Post-Vaccination and Viral Illness

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## ABSTRACT

Opsoclonus-myoclonus syndrome is a rare condition with dyskinesia of eye movements and myoclonic movements of the trunk and limbs. It is linked to malignancies, infections and other conditions. We present a case of post-vaccination opsoclonus-myoclonus syndrome. We believe that our patient also experienced a viral illness post-vaccination and prior to the development of opsoclonus-myoclonus. The vaccination in combination with the viral illness potentiated the immune mechanism thought to be noted in this condition. This was supported by her recovery after receiving immunotherapy.

**Keywords:** Opsoclonus; Myoclonus; Ataxia; Immunotherapy

## 1. Introduction

Opsoclonus is a dyskinesia of eye movements that persist during eye closure and sleep [1]. Opsoclonus is typically associated with myoclonic jerks of the trunk, limbs, and head giving rise to the name “opsoclonus-myoclonus syndrome (OMS)”. OMS is a rare disorder and is associated with infections or paraneoplastic mechanisms. Tumors commonly associated with OMS are small cell lung cancer, breast cancer, ovarian cancers [2] and neuroblastoma [3,4]. Other less common conditions related to OMS are HIV [5], toxins, pregnancy [1], and viral encephalopathic conditions (e.g. coxsackie virus, rubella and mumps) [6]. Recently, West Nile virus [7], Lyme disease [8], and Varicella-zoster virus [9] have also been linked. In this case, the possible etiology of influenza vaccine with subsequent flu like illness causing an immune-mediated response triggering the OMS and its treatment is discussed.

## 2. Case Report

The patient is a 30-year-old woman who noted an acute onset of dizziness two weeks prior to presentation to neurology. She began to have nausea and vomiting a few days after the onset of dizziness. Subsequently, she noted unsteady gait and jerkiness of eye movements bilaterally. Of note, she had received the flu vaccination two weeks prior to the onset of her symptoms. She also reported mild, self limited flu like symptoms three days after receiving the flu shot. Her past medical history, family and

social history were unremarkable. She was not on any medications. On examination, she was alert and showed normal mentation. Cranial nerve examination was intact except for opsoclonus. On motor examination, spontaneous myoclonus was noted throughout her body, which was exaggerated by active movement. She had full strength and intact reflexes symmetrically. She was unable to stand independently secondary to severe truncal ataxia. She also showed significant appendicular ataxia during finger to nose testing. Her sensory exam was intact to all modalities.

Work up included extensive blood test and spinal fluid analysis which were unremarkable and summarized in **Tables 1** and **2**. MRI of the brain with and without contrast was normal. CT scans of chest, pelvis, and abdomen were unremarkable. Mammogram was also unremarkable.

She was diagnosed with idiopathic OMS. In this case, the onset of her OMS was thought to be related to the influenza vaccination and possible viral infection acting as a second insult. Symptomatic treatment with clonazepam and levetiracetam was unhelpful. She was given intravenous methylprednisolone (1 g/d) for 3 days which showed minimal improvement. Plasmapheresis was initiated. She showed improvement after 6 exchanges. Upon discharge she had persistent saccomania but only on upgaze, continued myoclonic jerking of the head, and a kinetic tremor bilaterally in her upper extremities. She continued with weekly plasma exchanges for six weeks as an outpatient. Five months after her last plasma exchange,

**Table 1. CSF studies.**

CSF	Results
Gram Smear	No organisms, +1 WBC
Culture	No growth 3 days
Nucleated cells	6 cells
Red blood cells	2 cells
Glucose	57
Protein	52
VDRL	Non-reactive
Lactate	1.2
LDH	238
EBV by PCR	Negative
CMV IgM	Negative
West Nile Virus: IgM, PCR, culture	Negative
Lyme Disease PCR	Negative
Cryptococcal Antigen	Negative
VZV by PCR	Negative
Paraneoplastic Autoantibodies Panel	
ANNA-1/anti-Hu	Negative at <1:2
ANNA-2/anti-Ri	Negative at <1:2
ANNA-3	Negative at <1:2
AGNA-1	Negative at <1:2
PCA-1/anti-Yo	Negative at <1:2
PCA-2	Negative at <1:2
PCA-Tr	Negative at <1:2
Amphiphysin Ab	Negative at <1:2
CRMP-5-IgG/anti-CV2	Negative at <1:2

**Table 2. CSF studies.**

Serum	Results
HIV	Negative
T. Whipplei	Negative
Endomysial IgA antibody	<3 (Negative)
Lyme IgM/IgG antibodies	Negative
ANA	
Anti-SS-A	Negative
Anti-SS-B	Negative
Anti-Smith	Negative
Anti-Jo-1	Negative
Scl-70	Negative
Centromere B	Negative
dsDNA	Negative
Histone	Negative
Paraneoplastic Antibodies	
Anti-Hu	Negative
Anti-Yo	Negative
Anti-Ri	Negative
Serum protein electrophoresis	Normal
Immunofixation of serum	Normal polyclonal distribution of immunoglobulins

the patient's neurological exam showed minimal abnormality.

### 3. Discussion

The immunopathogenesis of OMS is poorly understood. There appears to be humoral and cell-mediated immune mechanisms involved both in paraneoplastic and idiopathic syndromes [1]. In many cases the symptoms are reversible after treatment with immunotherapy [1,2,10]. In idiopathic OMS cases, most patients are seronegative for antineuronal antibodies, as in this case. Our patient received the influenza vaccine two weeks prior the onset of neurological symptoms. Given that humoral immune mechanisms seem to play a role in OMS, it could be possible that the vaccine or a combination of vaccine and mild viral infection generated an immune response that lead to antibodies causing neuronal dysfunction. There are other case reports of post-vaccine as well as post viral OMS. One case describes OMS after receiving the HPV vaccination [11] and another following Rubella vaccination [12]. Bhatia et al described a patient with only myoclonus two weeks after symptoms of influenza [13]. Our patient also developed symptoms around two weeks of receiving flu vaccination. However, our patient also developed a mild flu like illness around the time of vaccination the fact that this patient improved with plasmapheresis provides additional support for an immune related response causing OMS.

### 4. Conclusion

In general the clinical course of idiopathic OMS syndrome in adults has a good recovery accelerated by the use of immunotherapy [1,2,10]. Immunotherapy should be initiated along with a complete work up for underlying malignancy, infectious, systemic diseases and toxic etiology. The vaccination in combination with the viral illness potentiated the immune mechanism thought to be noted in this condition. This was supported by her recovery after receiving immunotherapy.

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