

## A rare case of chickenpox induced myoclonus

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### Abstract

In the presence of abnormal neurological features, infective etiology should be kept as one of the differential diagnoses. This case report is about a 38 years old male patient who presented with fever with blister-like rashes in centripetal distribution over the body and myoclonus. Cerebrospinal fluid examination showed the presence of varicella-zoster IgM antibodies and a diagnosis of chickenpox induced myoclonus was made. Appropriate treatment recovered the patient completely. This case report highlighted the clinical spectrum of chickenpox as well as the possible pathogenesis and diagnostic, therapeutic approach of this uncommon entity.

**Key words:** Chickenpox, Myoclonus, Varicella-zoster.

### Introduction

Chickenpox is a highly contagious disease caused by the varicella-zoster virus (VZV). It is characterized by itchy, blister-like rashes having centripetal distribution over the body. Diagnosis is mainly clinical. Known complications were pneumonia, encephalitis, cerebellar ataxia, septicemia. Myoclonus is the sudden, involuntary, jerky contraction of muscle or group of muscle.<sup>1-5</sup> The syndrome of isolated myoclonus following infection has not been clearly defined. Myoclonus has been described in acute encephalitis due to a variety of viral and non-viral agents which either directly invade the CNS or by an immunological process.<sup>1</sup> The isolated post-infectious myoclonus is appropriate to describe the myoclonic syndromes that satisfy the following criteria: 1) A sudden

onset of generalized, multifocal, or segmental myoclonus, 2) a history of a recent preceding infectious illness, 3) no features of encephalitis or the opsoclonus myoclonus syndrome, 4) a non-progressive course without seizures, ataxia, or dementia and 5) recovery in a short but variable period of time.<sup>1</sup> This case report highlights the unusual presentation of chickenpox as well as the diagnostic and therapeutic approach of the entity.

### Case report

This case report is about a 38 years old Hindu male patient presented in April 2019. He was a resident of the Dungarpur district, Rajasthan, India and he was a jeweler by occupation. He presented with a history of fever 8 days, rashes over the body for the last 6 days, and abnormal jerky movement of the body for 3 days. A detailed history was taken. Fever was moderate, non-documented, intermittent, associated with generalized weakness. Rashes were insidious onset, gradually progressive. Rashes first appeared on the chest then involved other body parts in the sequence of the abdomen, back, face, upper, lower limb within 2 days. Rashes had blister-like projection, oozing watery discharge after the burst and later dried up with the appearance of new similar lesions. Rashes were denser over the trunk than face and extremities, associated with itching. After 5 days of onset of fever, the patient developed abnormal jerky involuntary movements of the trunk and limbs. The patient was having difficulty in daily routine activity as well as it was difficult to sleep properly due to abnormal jerky movements, although disappearance of jerks was there during sleep. On general physical examination vitals like blood pressure (128/84 mm Hg), pulse (84/minute, regular), respiratory rate (16/minute) were within normal limits. There were blisters like red rashes all over the body

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of varying sizes from 1 to 3mm. Rashes were denser over the trunk than face and extremities, sparing palm and sole. Few rashes were having intact vesicles, few blisters over rashes were oozing watery discharge and few rashes were crusted and dried up. No, another significant abnormality was found on general physical and systemic examination besides abnormal jerky movements of the trunk with limbs with frequency varying from 8 to 13 per minute. (Video 1 <https://youtu.be/wLaIZze9Aq0> and Video 2 <https://youtu.be/mfj55XVLPaw>)

The clinical diagnosis of chickenpox with myoclonus was made due to the presence of fever with typical blister-like rashes in centripetal distribution, in different stages of development and there was positive contact history with a chickenpox patient also. The patient was admitted to a proper isolation facility.

Previous literature was reviewed and the patient was investigated accordingly. A fever profile was sent to exclude other infections. Malaria antigen detection test and peripheral blood film examination were normal. Dengue NS1 antigen, Dengue IgM, IgG antibodies, Typhoid IgM, IgG antibodies, Scrub typhus IgM, IgG antibodies, VDRL were negative. Urine examination was also normal. Routine investigations had hemoglobin 13.6 mg/dl, total leukocyte counts 10,100/mm<sup>3</sup>, neutrophils 72%, lymphocytes 15.3%, monocytes 8.2%, eosinophils 2.1%, platelet count 213000/mm<sup>3</sup>, random blood sugar 113 mg/dl. Renal and liver function tests were also within normal limits. No significant abnormality was found in ECG, chest X-ray, and ultrasonography of the abdomen. He was further investigated by cerebrospinal fluid (CSF) examination and MRI brain. CSF examination showed cells 4 cells/mm<sup>3</sup>, glucose 60 nmol/L, protein 33 g/L, and varicella-zoster IgM antibodies were positive in the CSF. MRI brain was normal.

The patient was treated with injection acyclovir 500 mg IV 8 hourly, Injection dexamethasone 6 mg IV 8 hourly, intravenous fluid in the form of normal saline to maintain fluid balance and ensure proper hydration, tablet cetirizine 10 mg once a day, tablet pantoprazole 40 mg once a day, tablet baclofen 10 mg twice a day, tablet clonazepam 0.5 mg once a day at night, tablet paracetamol 500mg as per need. The patient was recovered as myoclonus was subsided within 3 days, fever subsided after 4 days and all rashes were crusted and dried up after 5 days. The patient was discharged after 5 days with treatment of tablet acyclovir 400 mg 8 hourly, tablet methylprednisolone 16 mg once a day for 5 days followed by 8 mg once a day for next 5 days, tablet cetirizine 10 mg once a day, tablet pantoprazole 40 mg once a day, tablet clonazepam 0.5 mg once a day at night, tablet paracetamol 500mg as per need. On follow up the patient was recovered completely in the next 10 days. No further signs and symptoms appeared in the next six months follow up without any treatment.

## Discussion

This article reported an uncommon presentation of chickenpox as myoclonus. The case report also highlighted that early diagnosis and appropriate treatment may do a complete recovery. There were only a few case reports about uncommon neurological presentations of chickenpox like- isolated post-infectious myoclonus,<sup>1</sup> cerebral ataxia,<sup>2</sup> opsoclonus-myoclonus syndrome,<sup>3</sup> optic neuritis with encephalitis,<sup>4</sup> opsoclonus-myoclonus-ataxia syndrome.<sup>5</sup> Opsoclonus is rapid conjugate multidirectional eye movements. Myoclonus is defined as sudden, involuntary, jerky contraction of muscle or group of muscle.<sup>5</sup>

Like the current study, Bhatia K et al<sup>1</sup> reported two cases of isolated post-infectious myoclonus. The study suggested that EEG response may indicate the cortical or not cortical origin of myoclonus. The study also mentioned that on the literature review, no EEG correlation was found with myoclonic jerks.<sup>1</sup> Both the cases of Bhatia K et al<sup>1</sup> had normal brain imaging like in the current case study and one case had normal CSF findings. Another case in the report refused a CSF examination. The study did not demonstrate infection of the neurological system. The study also did not mention the treatment approach of the patients. Both the cases were of post infectious myoclonus but in the current case report myoclonus were present during active infection, suggest infectious and/ or related immunological insult as etiology of myoclonus.

The current case also had normal brain imaging and CSF findings. The varicella-zoster specific antibodies were also demonstrated in the CSF in the current study. In the current study, antiviral antibiotics along with steroids were used. Steroids were used due to possible immunological injury to the nervous system, manifesting myoclonus. No focal lesion on brain imaging, generalized involvement of myoclonus, and presence of IgM antibodies in CSF also favored the immunological possibility. The recovery of the patient was also significantly faster than the previous case report by Bhatia K et al<sup>1</sup> (few days versus a few months). Fast recovery by antiviral antibiotic and steroid favored infective as well as immunological pathogenesis. Thus current study highlighted possible immunological injury to the nervous system and the role of steroids in the treatment. Further comparative studies may need to establish steroids as a treatment modality.

A study by Singh D<sup>3</sup> demonstrated varicella-zoster IgM antibodies in serum and CSF, in a patient with Opsoclonus-myoclonus syndrome. The patient also had pleocytosis predominantly lymphocytes with normal protein and sugar levels in CSF. The patient was treated with clonazepam due to its interaction with benzodiazepine receptors in the brain that facilitate inhibitory GABAergic transmission. In the study, steroid was not used due to possibility

of flaring of infection. Although the study mentioned that corticosteroids, intravenous immunoglobulins, immunosuppressants, plasmapheresis, rituximab, ACTH, clonazepam, baclofen, valproate, 5-hydroxytryptophan can be the useful modality of treatment. In that study, the opsoclonus and myoclonus completely disappeared over the period of 15 days. The current study used clonazepam along with baclofen, antiviral antibiotics, and steroids for treatment. The recovery of the patient was much faster than other studies as myoclonus subsided within 3 days only.

### Conclusion

Chickenpox may induce myoclonus. Early diagnosis and appropriate treatment may do complete recovery of signs and symptoms like in the current study. The use of steroids as treatment was not clearly mentioned in previous studies. In this case-report steroid was used due to the possibility of immunological etio-pathogenesis. Generalized involvement, no focal lesion on imaging, and presence of antibodies in CSF also favored this fact. In the current study recovery of the patient was faster than in previous studies. This indicates steroids can be a useful modality of treatment. Further studies are needed to establish steroids role. Antiviral antibiotics, baclofen, clonazepam, and proper hydration are also useful in treatment.

**Conflict of Interest:** None

**Source(s) of support:** None

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