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Clinical Short Communication

Opsoclonus myoclonus ataxia associated with West Nile virus infection: A dramatic presentation with benign prognosis?

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ABSTRACT

Opsoclonus myoclonus and ataxia is a combination of severe neurological signs associated with several pathologic agents and conditions. Only few cases of opsoclonus have been related to West Nile virus infection. We report on a 61-year-old woman and on a 55-year-old man who had history of recent fever, who were hospitalized because of acute severe truncal ataxia, opsoclonus and tremor with minimal myoclonic jerks. A through work-up revealed the presence of both IgM and IgG antibodies against West Nile virus both in the serum and Cerebrospinal Fluid and excluded other causes known to be associated with this combination of neurological signs. The first case was treated with corticosteroids, followed by significant improvement, and the second recovered spontaneously. The acute combination of opsoclonus, severe truncal ataxia and tremor with a history of recent fever requires, during the relevant season and in the relevant geographic area, a search for a recent infection with West Nile virus. Though initially suffering from a devastating sickness, our patients eventually recovered.

1. Introduction

Opsoclonus is a rare phenomenon of combined multidirectional, involuntary, arrhythmic and chaotic saccadic oscillations with a frequency of 10 to 15 per second, without an intersaccadic interval [1,2]. Although infectious and post-infectious etiology has been well recognized; we are aware of only six case reports of opsoclonus related to West Nile infection [3–8]. We present two cases of severe opsoclonus-myoclonus-ataxia syndrome (OMAS) associated with West Nile virus (WNV) infection.

2. Case 1

A 61-year-old woman with a history of diabetes mellitus, hypertension, hyperlipidemia and obesity was admitted with two weeks history of fever, nausea and vomiting. Five days prior to admission she started complaining of progressive dizziness, imbalance and difficulty focusing. On admission neurological examination revealed severe opsoclonus, truncal ataxia (not able to ambulate) and tremor mainly of the head, and less of the hands with minimal myoclonus (Video clip). Chest X-ray was normal and routine blood tests were normal. Brain MRI showed evidence of mild microangiopathic lesions probably related to her diabetes mellitus and hypertension (Fig. 1). Lumbar puncture yielded 13 mononuclear leukocytes/mm^3 and normal levels of protein and glucose in the CSF (cultures were negative for bacteria and candida). Blood and urine cultures were unremarkable. Paraneoplastic antibodies, anti-Yo and anti-Ri, were not detected in the blood. Levels of antithyroid, antiphospholipids, antinuclear and antineutrophil cytoplasmic antibodies were within normal limits. Computed tomography of the chest, abdomen and pelvic did not demonstrated significant findings. Mammography and gynecology examination were normal. After receiving the CSF results, intravenous methylprednisolone 1 g per day was given for a week with significant improvement. Later, results of WNV serology came back: IgM and IgG antibodies were detected in CSF and blood, suggesting recent infection. All symptoms resolved over three months.

3. Case 2

A 55-year-old man was admitted to the neurology department with a history of fever and general weakness starting two weeks previously. This was followed four days later by vomiting, dizziness and falling. On admission, he was irritable, not able to walk, and on examination he had action tremor with minimal myoclonic jerks, severe truncal ataxia and primarily opsoclonus followed by ocular flutter (repetitive, irregular, involuntary bursts of horizontal saccades without an inter-saccadic interval - Video clip). Chest X-ray was normal and routine blood tests showed mild leukocytosis. Magnetic resonance imaging (MRI) of the brain demonstrated an old small right thalamic infarct (Fig. 2). A lumbar

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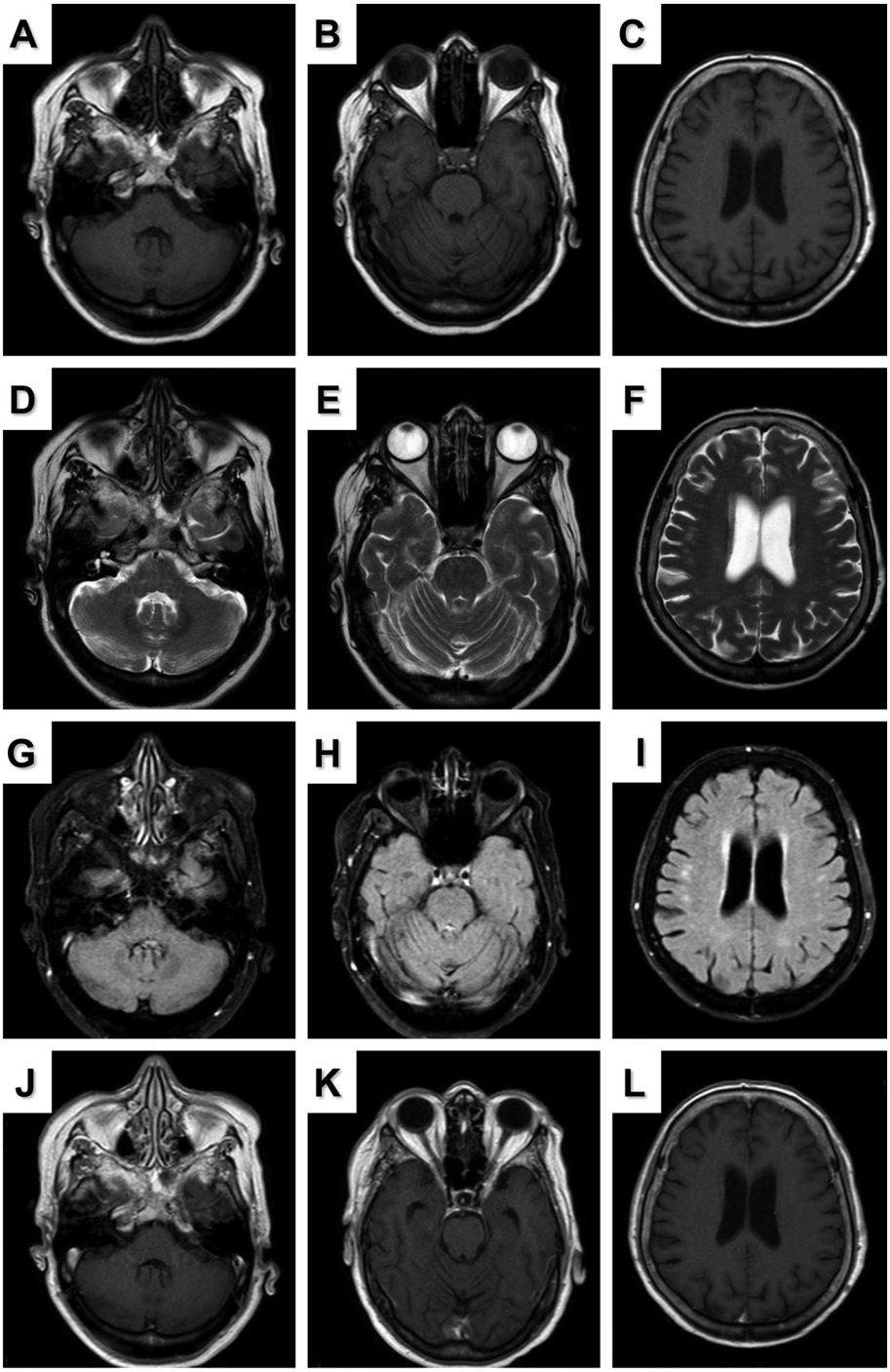

Fig. 1. Patient 1. Axial T1 weighted (A–C), T2 weighted (D–F), Fluid attenuated inversion recovery - FLAIR (G–I) and T1 weighted with gadolinium contrast (J–L) MRI images showing only mild microangiopathic changes (F and I images).

puncture was performed; no cells were found with normal glucose level (55 mg/dl, normal range; 50–80 mg/dl), but the protein concentration was high (88 mg/dl, normal range: 15–50 mg/dl). Polymerase chain reaction did not detect a DNA fragment of Herpes Simplex virus in the CSF, and CSF cultures were negative for bacteria and candida. Blood and urine cultures were unremarkable. Levels of antithyroid, antiphospholipids, antinuclear and antineutrophil cytoplasmic antibodies were within normal limits. Paraneoplastic antibodies, anti-Yo and anti-Ri, were not detected in the blood. Computed tomography of the chest, abdomen and pelvic did not demonstrated significant findings. IgM and IgG antibodies against WNV were detected in CSF and blood, suggesting recent infection. All symptoms progressively improved and resolved over a few months.

4. Discussion

The etiology of opsoclonus may be paraneoplastic, post infectious, infectious, metabolic, toxic or miscellaneous [1]. Several immune mechanisms have been proposed, involving both the humoral immune system (anti-Ri, anti-Yo, anti-Hu and others antibodies have been associated with the phenomenon) and cell mediated immune mechanisms [1].