Aseptic Meningoencephalitis Presenting with Bilateral Vestibular Ataxia: a Case Report

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Abstract

Bilateral vestibular dysfunction is a rare condition, of which peripheral disorders are most common, whereas central disorders are extremely rare. A 35-year-old woman developed fever, headache, dizziness, convulsion, and disturbance of consciousness at the same time. MRI findings were normal. Based on the neurological findings and laboratory abnormalities, she was diagnosed as having aseptic meningoencephalitis. Steroid pulse therapy successfully ameliorated her encephalitis sign. However, isolated bilateral vestibular ataxia and dizziness, together with severe decrease of Caloric nystagmus, became apparent and lasted for the following 3 months, without cerebellar/brainstem involvement.

Key words: bilateral vestibular ataxia, aseptic meningoencephalitis, dizziness, vestibular dysfunction

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Introduction

Bilateral vestibular dysfunction is a rare but well-documented condition. Peripheral disorders are the most common aetiologies (e.g., gentamicin ototoxicity, otological/neoplastic diseases, or cranial/peripheral neuropathies), which are often accompanied by hearing loss (1). In contrast, central disorders [e.g., spinocerebellar degeneration (2), meningeal carcinomatosis (3), or meningitis (4)] rarely cause such condition. We recently had a patient who presented with aseptic meningoencephalitis and bilateral vestibular ataxia together.

Case Report

A 35-year-old, previously healthy woman began to have fever, headache, and dizziness. Two days later, she developed tonic convulsion. On her referral to our hospital, she was drowsy and disoriented. She had a stiff neck, positive Kernig sign, and extensor plantar responses. Laboratory examination showed a mild inflammatory reaction alone, and systemic vasculitis and anti-phospholipid disorder were excluded. The cerebrospinal fluid (CSF) examination showed

an increase in cell count (237/mm³; polymorphonuclear, 32; mononuclear, 205) and total protein (77 mg/dl) and a mild decrease in glucose content (45% of serum glucose). Brain magnetic resonance imaging (MRI) scans were normal. Electroencephalography showed a diffuse, slowed, and irregular rhythm. The neurological findings and the laboratory abnormalities were consistent with the diagnosis of aseptic meningoencephalitis. We initially suspected the cause of meningoencephalitis was viral, and started 1500 mg/day of acyclovir, 600 ml/day of glycerol, 24 mg/day of dexamethasone, and 100 mg/day of phenobarbital, without antibiotics such as aminoglycosides. These treatments ameliorated her symptoms mildly. However, they exacerbated again along with tapering of dexamethasone, whereas we kept the same dose of acyclovir. At that time, results of bacterial culture and viral studies (Ebstein-Bar, varicella-zoster, cytomegalovirus, herpes simplex, coxsackie A16, and enterovirus) turned out negative by enzyme immunoassay and polymerase chain reaction. At the eighth hospital day, we stopped acyclovir, and started two courses of 1000 mg/day, 3 days of intravenous methylprednisolone, which was followed by 60 mg/day of oral prednisolone. Two days after starting methylprednisolone, disturbance of consciousness disappeared completely. When she resumed sitting up in

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bed, however, she noticed by herself severe unsteadiness in sitting/standing, and dizziness particularly when she was looking around. On examination, she had no cranial nerve involvement (except for the vestibular system), no cerebellar ataxia in speech, extra-ocular movement, or limb coordination, or decreased sensation for position and vibration. However, she had unsteady, wide-based gait, with positive Romberg sign without laterality. Results of audiogram, auditory evoked potential, and eye tracking test were normal. However, caloric-evoked nystagmus was severely decreased bilaterally, and a postulography showed a peak body-sway frequency around 1 Hz (1 Hz, deep sensory/vestibular; 3 Hz, cerebellar) and an increased Romberg quotient. All these findings suggested bilateral vestibular ataxia of peripheral nerve/end-organ origin. Three months later, along with gait rehabilitation, oral prednisolone was gradually tapered, and the patient's gait ataxia and dizziness disappeared almost completely.

Discussion

In the present patient, the CSF examination showed no evidence of bacterial or viral infection, although we could not completely exclude virus as a cause of aseptic meningoencephalitis. Steroid pulse therapy rapidly ameliorated her encephalitic symptoms (5). This was most probably brought

about by the anti-edema and anti-inflammatory actions of corticosteroid. It is remarkable that bilateral vestibular ataxia occurred together with aseptic meningoencephalitis in our patient, which might have peripheral nerve/end-organ origin. Neurological, neuro-otological, and MRI findings did not show any evidence of cerebellar or brainstem involvement in the patient. Steroid pulse therapy ameliorated the bilateral vestibular ataxia slowly. Previously, a combination of bilateral vestibular dysfunction and aseptic/viral meningitis has only rarely been reported, and the mechanisms that underlie the condition remain obscure (1, 6, 7). Recently, autoimmune bilateral vestibulopathy has been reported (8). It usually occurs together with sensory deafness (9), but may precede sensory deafness or occur alone (8, 9). In such cases, antibodies to inner ear antigens were demonstrated. Seventy percent of the cases treated with corticosteroids improved clinically, often in a short term, but the remaining cases were resistant to aggressive immunosuppression (9). The occurrence of bilateral vestibular dysfunction and aseptic meningoencephalitis in our case suggested a possibility that such auto-antibodies might contribute to her gait ataxia and dizziness. Further cases are awaited to clarify the underlying mechanism. In conclusion, a rare combination of aseptic meningoencephalitis and bilateral vestibular ataxia was reported. Further cases are awaited to clarify the understanding mechanism.

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