Case Report

Morvan's syndrome with anti contactin associated protein like 2 – voltage gated potassium channel antibody presenting with syndrome of inappropriate antidiuretic hormone secretion

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ABSTRACT

Morvan's syndrome is a rare autoimmune disorder characterized by triad of peripheral nerve hyperexcitability, autonomic dysfunction, and central nervous system symptoms. Antibodies against contactin-associated protein-like 2 (CASPR2), a subtype of voltage-gated potassium channel (VGKC) complex, are found in a significant proportion of patients with Morvan's syndrome and are thought to play a key role in peripheral as well as central clinical manifestations. We report a patient of Morvan's syndrome with positive CASPR2-anti-VGKC antibody having syndrome of inappropriate antidiuretic hormone as a cause of persistent hyponatremia.

Key words: Anti-voltage-gated potassium channel antibodies, hyponatremia, myokymia, Morvan's syndrome, syndrome of inappropriate antidiuretic hormone

Introduction

Morvan's syndrome is a rare autoimmune disorder characterized by peripheral nerve hyperexcitability, autonomic dysfunction, and central nervous system symptoms.^[1] The syndrome of muscle twitching, dysautonomia, insomnia, and delirium was first reported by Morvan by the name of la choree fibrillare in 1890.^[2] We report a rare case of contactin-associated protein-like 2 (CASPR2), a subtype of voltage-gated potassium channel (VGKC) complex antibody positive Morvan's syndrome, with syndrome of inappropriate antidiuretic hormone secretion (SIADH).

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Case Report

A 45-year-old male presented with 4-month duration of nonradiating mild back pain, followed a month later by burning sensation in palms and soles with nocturnal exacerbations. He developed abnormal twitching of muscles in both upper and lower limbs. He became aggressive, over-talkative, and insomniac over 15 days before presentation. He had significant weight loss during the period.

On examination, he was anxious and restless, having resting tachycardia and excessive sweating. His higher mental function and cranial nerves examinations were normal. He had continuous undulating twitching in both upper and lower limbs and back muscles. His knee and ankle jerks were sluggish and rest of the examination was normal.

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Hemogram, renal, liver, and thyroid functions were normal. Electromyography showed spontaneous activity including myokymic discharges [Figure 1], doublets, and triplets in both upper and lower limb muscles. Magnetic resonance imaging of the brain and lumbosacral spine was normal. The patient had positive serum anti-CASPR2 antibody, a subtype of VGKC complex detected by immunofluorescence method. His cerebrospinal fluid examination showed raised proteins 76 mg/dl (normal: 20–40 mg/dl), with normal cell count (cells: 3/mm³, all lymphocytes). Computed tomography (CT) of the chest showed no evidence of thymoma. The patient was diagnosed as Morvan's syndrome with positive anti-CASPR2 – VGKC antibody.

There was persistent low serum sodium in the range of 125–130 mEq/L, for which patient was evaluated. His urinary osmolality was raised (216.36 mOsm/kg, normal <100 mOsm/kg) and random urinary sodium was increased (42 mmol/L, normal <30 mmol/L). The serum osmolality was decreased (271.5 mOsm/kg) and urinary specific gravity was 1.010. These findings showed SIADH secretion as a cause of his persistent hyponatremia. The patient was treated with intravenous immunoglobulin (IV Ig) 2 g/kg in 5 divided doses. He was given phenytoin at dose of 100 mg three times a day for symptomatic relief for twitching, which acts as membrane stabilizer. The patient was started on oral prednisone (1 mg/kg) and fluid restriction was advised.

He had marked improvement in muscle twitching and was able to sleep properly with immunotherapy. Electromyography done 2 weeks after the course of IV Ig showed decrease in spontaneous activity; occasional fasciculations were seen. His hyponatremia was also corrected. On follow-up, after 3 months, the patient was completely normal and electromyography showed no spontaneous activity. Oral prednisone was given

1 mg/kg for 3 months and later tapered gradually over next 2 months.

Discussion

Morvan's syndrome is characterized by myokymia associated with muscle pain, excessive sweating, weight loss, hallucinations, sleep disorders, and behavioral abnormality.^[1,3] This is considered as a form of neuromyotonia having prominent central features. There was considerable overlap between central and peripheral features in our patient. The basic mechanism for both presentations is same, that is anti-VGKC antibody, acts at different levels of neuraxis, both at central and peripheral level.^[4] Association of SIADH is uncommon in VGKC–CASPR2 antibodies positive cases but common in anti-leucine-rich glioma inactivated-1 (LGI-1) antibodies positive cases.^[5]

VGKC-complex antibodies include both CASPR2 and LGI-1 antibodies. LGI-1 antibodies are usually associated with hyponatremia, and CASPR2 antibodies are usually associated with thymomas which carry poor prognosis. CASPR2 antibodies mostly bind the neuropil, whereas antibodies to LGI-1 bound to neuronal cell bodies including the antidiuretic hormone-secreting and orexin-secreting hypothalamic neurons present in hypothalamus, raphe nucleus, and locus coeruleus. Hyponatremia is not commonly reported in Morvan's syndrome although it is present in half of the patient in LGI-1 antibodies positive limbic encephalitis.^[5]

The classical electromyographic finding is the myokymic and neuromyotonic discharges. In addition, fasciculation, doublets, triplets, multiplets, and positive sharp waves are also present. Management includes antiepileptic drugs and immunotherapy which improve the clinical and electrophysiologic manifestations.^[6]

This is a rare case report of Morvan's syndrome with SIADH having anti-CASPR2–VGKC antibodies that responded to immunosuppression.

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Conflicts of interest

There are no conflicts of interest.

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Figure 1: Spontaneous activity in right tibialis anterior showing myokymic discharges (sweep speed: 0.1 ms and sensitivity: 50 µV)

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