ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 05 | May - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

MORVAN'S SYNDROME : A RARE CASE REPORT

General Medicine

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ABSTRACT

Morvan's syndrome is an autoimmune disorder characterized by central, autonomic, and peripheral hyperactivity with associated thymoma in majority. The clinical features include insomnia, hyperhidrosis, myokymia, dysautonomia and hallucinations. Voltage gated potassium channel (VGKC) namely, leucine-rich glioma inactivated 1 (LGI-1) antibody and contactin-associated protein-like 2 (CASPR-2) antibody are thought to play a pathogenic role in peripheral and central nervous system symptoms in Morvan's syndrome. Electromyography (EMG) is useful in diagnosis of Morvan's syndrome characterized by spontaneous myofiber activity with various denervation potentials. Computed tomography (CT) scan and Magnetic resonance imaging (MRI) brain is usually normal. Immunosuppressive therapies, encompassing corticosteroids, azathioprine, methotrexate and recently, rituximab, are the mainstay of therapy. Other treatments include PE, IVIG, and thymectomy. We report a rare case of morvan syndrome with typical clinical features along with leucine-rich glioma inactivated 1 (LGI-1) antibody and contactin-associated protein-like 2 (CASPR-2) antibody positivity successfully treated with intravenous immunoglobulin (IVIG) therapy.

KEYWORDS

Morvan's syndrome, VGKC antibodies, myokymia

INTRODUCTION:

The French physician Augustine Marie Morvan described a syndrome consisting of muscle twitching, dysautonomia, insomnia and fluctuating delirium by the name 'la chorée fibrillaire de Morvan' in the late 19th century(1). It is now known as Morvan's syndrome. We report a rare case of morvan syndrome with typical clinical features along with leucine-rich glioma inactivated 1 (LGI-1) antibody and contactin-associated protein-like 2 (CASPR-2) antibody positivity.

Case Report:

A 28 year old female patient presented with hyperhidrosis since 20 days, insomnia since 15 days, bilateral limb pain since 10 days, twitching over the bilateral calf muscles 4- 5 times/ day since 7 days, for which she was admitted at a tertiary care hospital. On 2nd day of admission, patient developed myokymia of facial and ocular muscles.

General examination revealed Pulse rate of 124/min and Blood pressure 110/70 mm Hg. Spontaneous fasciculation over bilateral calf muscles. Neurological and cranial nerve examination were normal. Power was 5/5 in all four limbs. Sensory examination and tendon reflexes were normal.

Laboratory investigations were normal except hyponatremia. MRI of brain revealed no significant abnormity. Electrophysiological studies showed inelicitable F-responses over the bilateral tibial nerves. EMG study revealed fibrillation and fasciculations spontaneously in right and left soleus, suggestive of muscle membrane instability. CT of chest and abdomen was normal; no signs of thymoma were seen. The serum immunofluorescence was positive for LGI-1 and CASPR-2 antibodies (VGKC antibodies).

The diagnosis of Morvan's syndrome was made on clinical features typical EMG and positive LGI-1 and CASPR-2 antibodies. The patient was given trial of intravenous methylprednisolone pulse therapy for 3 days. Since there was no improvement patient was started on intravenous immunoglobulin (IVIG) 2g/kg over 5 days, followed by tapering doses of oral prednisolone 40mg x 10 days,30mg x 10 days,20mg x10 days, 10mg x 10 days, 5mg × 10 days, 2.5mg x 10 days.On fifth day of starting IVIG there was improvement in myokymia and twitching. After 7 days of intravenous immunoglobulin (IVIG) treatment all symptoms improved but there was persistence of limb pain for which patient was given pregabalin. After 1month follow up, lower limb pain also improved. At present patient is under remission and has resumed her job.

Patient was evaluated to rule out underlying malignancy with Radiological and Biochemical investigations.

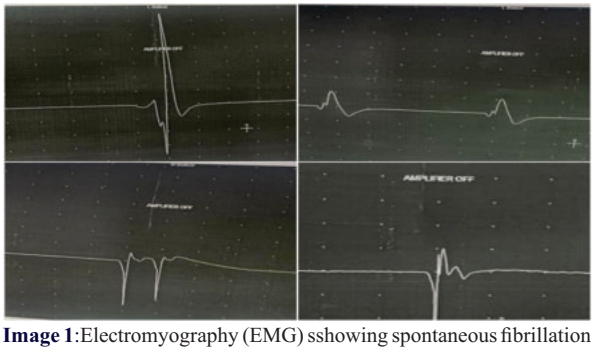

Image 1:Electromyography (EMG) sshowing spontaneous fibrillation in bilateral soleus muscle.

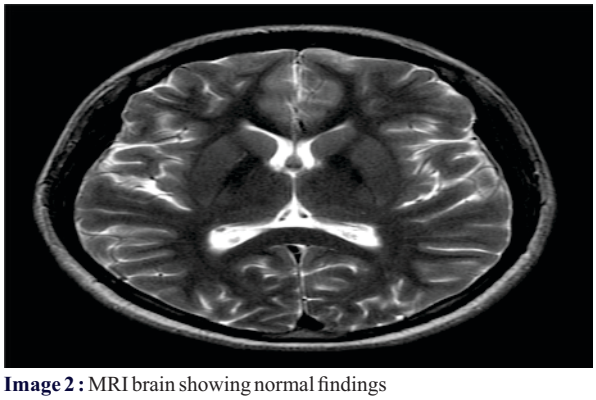

Image 2 : MRI brain showing normal findings

DISCUSSION:

Our case was female, while according the literature Morvan's syndrome has a male preponderance with a male to female ratio of 13:1 (2). Insomnia is a common CNS manifestation in Morvan's syndrome which was also reported in our patient (3).Similar to cases reported previously, MRI of brain was normal in our patient (4). VGKC antibodies are presumed to play role in pathogenesis of Morvan's syndrome and have been detected in 50% of patients (5). VGKC (both

CASPR-2 and LGI-1) antibody positivity was also seen in our patient. Our patient reported myokymic discharges on EMG as seen in patients with Morvan's syndrome (6).

CONCLUSION:

Morvan's syndrome with both LGI-1 and CASPR-2 antibody is reported rarely. With high index of suspicion this patient could be diagnosed and treated early.

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