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A Case of Neuronal Hyperexcitability Syndrome

Case Report

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Introduction

Morvan syndrome is an autoimmune disorder characterized by autoimmune antibodies directed against Voltage gated potassium channel (VGKC) [1]. Leading to peripheral nerve hyperexcitation causing muscle cramps, myokymia, fasciculation, neuropathic pain and dysautonomia. VGKC are transmembrane channels complexed with various proteins, such as leucine-rich glioma inactivated protein 1 (LGI1), contact in-associated protein 2 (CASPR2) and contactin [2]. Morvans syndrome can be post infectious, autoimmune, paraneoplastic or could be aggravated by toxins too.

Case Report

13 years female presented with painful crampsand twitching in bilateral forearms, hands, thighs, and soles along with excessive irritability and decreased sleep she was apparently normal till 3 months back then developed, Subacute onset progressive painful cramps involving back of thighs, forearms and both hands following a fever episode. Fever was of high grade lasted for three days, not associated with diarrhea, cough with expectoration, Headache altered sensorium. Pain progressively increased in intensity with tremors and palpitation. Patient also noticed Wide spread muscle twitching in both upper and lower limbs. She also gave history of excessive sweating in both palms and feet. Significant History of itching and peeling of skin in both hands and feet since 1 month. Her mother gave history of Excessive irritability and behavioral abnormality, decreased sleep, only 2 -3 hours of sleep per day. No history suggestive of weakness of limbs. No history suggestive of loss of pain touch sensation. No history suggestive of unsteadiness or difficulty in coordination. No history of altered bowel and bladder sensation. No history of difficulty in speech. No history suggestive of cranial nerve involvement. No history of LOC, Seizures, Trauma. General Examination Revealed Patient conscious alert, peeling of skin noticed in palms and soles, Hyperhidrosis noticed in palms and soles (Figure 1a, 1b, 2).

HMF examination showed normal status with a Mini Mental Status Examination (MMSE-30/30). Cranial nerve examination revealed no deficits. Motor examination of muscle tone, power was normal and all DTR are present. Sensory examination was

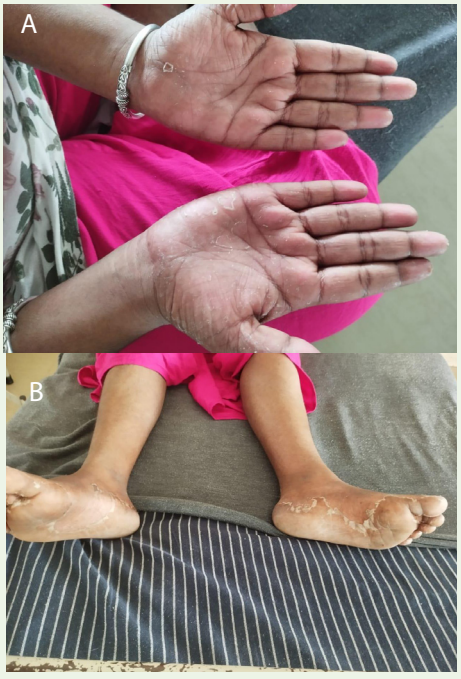

Figure 1A,B: Images showing peeling of skin

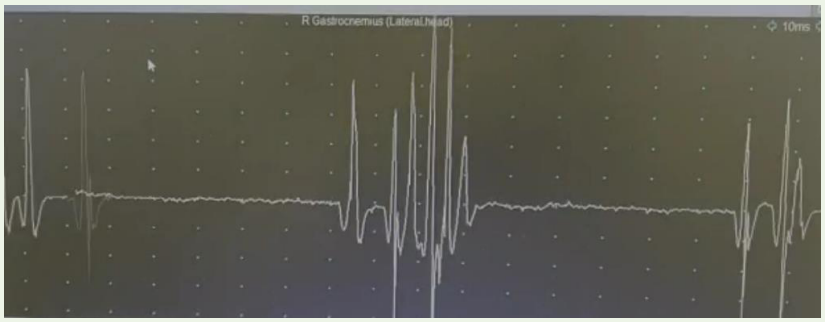

Figure 2: Image EMG showing myokymic potential

normal. Coordination normal. Fasciculation seen in trunk and limbs provisional diagnosis of neuromyotonia is made and case is worked up

EMG was done and it showed myokymic potentials with couplet and triplets.

ANA and ANCA – Negative

Antibody against Voltage Gated Potassium Channel (VGKC) revealed

Antibody to LGI 1-**Positive**

Antibody against CASPR - **Positive**

CT Chest and CT Abdomen done to rule out malignancy and revealed no malignancy.

MRI Brain and EEG was normal.

Treatment Given

IV steroids given for three days followed by

Intravenous Immunoglobulin 2g/kg body weight over five days.

Tab. Carbamazepine 200 mg BD

Symptoms relieved well following treatment and she was discharged.

Discussion

Autoimmune disorders directed against voltage gated potassium channel include Neuromyotonia, limbic encephalitis and Morvans syndrome.[3]

Morvan syndrome described by Auguste Morvan,the term "la chorıe fibrillare" was first used by the French physician Augustine Marie Morvan [4]. It is a rare autoimmune disorder with auto antibodies against voltage-gated potassium channels (VGKC)., primarily two kinds of potassium channel proteins, CASPR2 and LGI1 [5]. LGI1-Ab and CASPR2-Ab are associated with Morvans syndrome, LGI1-Ab is usually linked with limbic encephalitis (LE), faciobrachial dystonic seizures, hyponatremia, and REM sleep behaviour disorders [6]. Morvan syndrome serum are usually directed against LGI1, CASPR2, or both, but CASPR2-Ab dominates [7]. It has both CNS and PNS manifestation.CNS manifestation include- severe insomnia, irritability, encephalopathy, dysautonomia. PNS manifestation include-painful cramps, myokymia, fasciculations, neuropathic pain [8]. It may also present with systemic features such as weight loss, skin lesions or pruritus. LGI1 is also located particularly in the hippocampus and CASPR2 in the hippocampus and cerebellum [9]. In many cases Morvan syndrome is paraneoplastic, being especially associated with thymoma and small cell lung cancer but it can also occur in the absence of any cancer [10]. It can also be associated with myasthenia gravis.There was direct association with indigenous medicines, formulations based on herbs, heavy metals, and various colloids. Brain MRI and PET scan of Morvan syndrome are usually normal, while electroencephalogram (EEG) sometimes shows diffuse slow waves, occasional 4–6 Hz theta wave activity, and a typical rapid eye movement (REM) with absence of non-rapid eye movement sleep (NREM) stage [11]. Electromyography is useful in diagnosis of Morvan syndrome, which is characterized by spontaneous myofiber activity with various denervation potentials [12]. Usually remission occurs following immunotherapy, spontaneous remission is less likely even death can occur due to autonomic dysfunction in untreated cases.

Conclusion

Multidisciplinary approach is recommended for the management of such cases. Our case describes the importance of clinical examination and highlights the importance of pattern recognition in neurology.

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