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Case Report

Morvan syndrome: Unravelling the engimatic neurological disorder and its immunomodulatory management

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ABSTRACT

Syndrome is by with autonomic and involvement. Syndrome is to be an autoimmune disorder including an association with to voltage-gated potassium channel (Anti-Vgkc -Ab). Associated protein like 2, and leucine rich inactivated protein. These antibodies cause peripheral producing painful cramps, and central producing confusion, Hallucinations, and memory problem and autonomic producing, Fluctuation in blood pressure. There are several suggesting that morvan syndrome may be closely associated with drug intake leading to heavy metal toxicity. Here, we present a case with clinical symptoms suggestive of morvan syndrome following intake traditional medicine.

1. Background

Morvan syndrome is a rare disorder involving the central nervous system, peripheral nerve hyperexcitability, and autonomic dysfunction.[1] It was Augustin-Marie Morvan, a French physician, who first described this syndrome as "la choree fibrillaire de Morvan" in 1980. Morvan syndrome is thought to be an autoimmune disorder, including an association with antibodies against voltage-gated potassium channels (VGKC) and thymoma.[2] Morvan syndrome is typically characterized by neuromyotonia, sleep dysfunction, dysautonomia, and cognitive dysfunction. Peripheral nerve hyperexcitability is characterized by cramps, fasciculations, as well as neuromyotonic and myokymic discharges on needle electromyography.[3] There are several reports suggesting that Morvan syndrome may be closely associated with ayurvedic drug intake leading to heavy metal toxicity.[4] Mercury poisoning can present with a clinical picture similar to Morvan syndrome.[5] Here, we present a case with clinical symptoms suggestive of Morvan syndrome.

2. Case Presentation

A 35-year-old nulliparous female, an accountant with no co-morbidities who was undergoing native infertility treatment for the past two months, developed generalized itching, initially in the perianal region, followed by difficulty in initiating and maintaining sleep, third-degree auditory hallucination, and delusions in the form of persecution. She sought treatment at a nearby local psychiatric clinic but discontinued the medication after three to four days. She also experienced unsteadiness of gait for the past five days, with restlessness in her lower limbs, abnormal twitching movements involving the oral region and calf muscles, and pill-rolling tremors. Initially, a detailed central nervous system examination was not performed. The patient was put on antipsychotics, and after two days, she developed fever, altered sensorium, and passed high-colored urine – she was diagnosed and treated as

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having Neuroleptic Malignant Syndrome. To rule out CNS infection, autoimmune encephalitis CSF analysis, including AI profile and Gene Xpert profile, was done. After ruling out CNS infection, the patient was started on steroids, and gradual improvement was noticed within 7-10 days. A detailed CNS examination later revealed acalculia, right to left confusion with bilateral cerebellar features. EEG revealed diffuse slow waves, and EMG showed neuromyotonic features.

2.1. Investigations

A comprehensive set of investigations, including complete blood count, renal function test, liver function test, creatinine phosphokinase, creatinine kinase, urine analysis, ANA profile, ENA profile, ESR, CRP, HCV, HIV, thyroid function test, peripheral smear, CSF analysis, blood culture, and sensitivity, urine culture, and sensitivity were done.

CSF analysis revealed anti-VGKC antibodies and elevated CSF protein.

A toxic profile for mercury was also obtained, and an evaluation for carcinoma was conducted. Radiological investigations, including CT scans of the brain, chest, abdomen and pelvis and MRI brain, were performed.

EEG and EMG were also done.

EMG: spontaneous activity - spontaneously occurring continuous, irregularly occurring discharges in doublet, triplet or multiple single motor unit discharges.

2.2. Differential diagnosis

The differential diagnoses considered included mercury toxicity, autoimmune encephalitis, and carcinoma.

Treatment initially, the patient was diagnosed with acute psychosis and treated accordingly by the psychiatric department with Olanzapine, Trihexiphenidyl, and Diazepam. However, two days later, the patient developed fever with altered sensorium, high-colored urine, and decreased urine output. She was then shifted to the Intensive Medical Care Unit, where she exhibited signs of increased tone in all four limbs, brisk deep tendon reflexes, and bilateral plantar withdrawal, with no signs of meningeal irritation. Suspecting autoimmune encephalitis, the patient was put on pulse steroid therapy, resulting in a tremendous improvement.

Outcome and Follow-up the patient showed a dramatic improvement symptomatically with steroid therapy, and she is now on maintenance therapy.

3. Discussion

Morvan syndrome is a rare and complex neurological disorder characterized by a unique combination of central nervous system involvement, peripheral nerve hyperexcitability, and autonomic dysfunction. This case report describes a 35-year-old female presenting with a constellation of symptoms suggestive of Morvan syndrome. The clinical manifestations included generalized itching, sleep disturbances, auditory hallucination, delusions, abnormal limb movements, and pill-rolling tremors. These symptoms initially led to a misdiagnosis of acute psychosis, and the patient was treated with antipsychotic medication. However, the subsequent development of fever, altered sensorium, and high-colored urine raised suspicion of a more complex neurological condition.[6]

The diagnostic process for Morvan syndrome involves a multidisciplinary approach. It is essential to consider autoimmune encephalitis, mercury toxicity, and neoplastic diseases in the differential diagnosis, as these conditions can present with overlapping clinical features. To rule out infectious causes, CSF analysis and other relevant tests were conducted, and autoimmune encephalitis was suspected, leading to the initiation of pulse steroid therapy, which resulted in significant improvement.

The key features of Morvan syndrome are peripheral nerve hyperexcitability, dysautonomia, and encephalopathy, often leading to severe insomnia and cognitive dysfunction. Electromyographic studies in this case revealed neuromyotonic features, further supporting the diagnosis. The presence of antibodies against voltage-gated potassium channels (VGKC) is commonly associated with Morvan syndrome, and in this case, the patient had positive VGKC antibodies, reinforcing the autoimmune nature of the disorder.[7]

Although Morvan syndrome is considered rare, it is essential for clinicians to be aware of this condition and its diverse clinical manifestations. Early recognition and prompt diagnosis are crucial as the disease can lead to significant morbidity and mortality if left untreated. The delayed diagnosis in this case illustrates the challenge faced by physicians in differentiating Morvan syndrome from other neurological and psychiatric conditions, emphasizing the importance of a thorough clinical evaluation.

The treatment approach for Morvan syndrome typically involves immunomodulatory therapies such as corticosteroids, intravenous immunoglobulins, and plasma exchange. In this case, pulse steroid therapy resulted in a remarkable improvement in the patient's symptoms. However, long-term management and follow-up are essential to monitor for disease relapse or complications.[8]

The etiology of Morvan syndrome remains unclear, and the association with thymoma and small cell lung cancer suggests a possible paraneoplastic origin in some cases. However, in this case, a neoplastic cause was excluded after a thorough evaluation. The potential role of ayurvedic drug intake leading to heavy metal toxicity as a trigger for Morvan syndrome was also considered, highlighting the importance of assessing potential environmental exposures in the diagnostic process.

In conclusion, this case report highlights the challenges in diagnosing Morvan syndrome due to its diverse clinical presentation and rarity. A multidisciplinary approach, careful evaluation of clinical features, and appropriate laboratory investigations are crucial for arriving at an accurate diagnosis. Prompt initiation of immunomodulatory therapy can lead to significant clinical improvement in patients with Morvan syndrome. Further research is needed to better understand the pathogenesis and management of this intriguing neurological disorder.

4. Source of Funding

None.

5. Conflict of Interest

None.

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