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CASPR2-ASSOCIATED LIMBIC ENCEPHALITIS WITH FEATURES OF MORVAN SYNDROME: A CASE REPORT

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ABSTRACT
Morvan syndrome is a rare autoimmune neurological disorder characterized by neuromyotonia, autonomic dysfunction, and central nervous system involvement, often associated with CASPR2 antibodies. We present a 77-year-old male with CASPR2-associated autoimmune limbic encephalitis and facial-brachial dystonic seizures (FBDS), who exhibited seizures, autonomic dysfunction, and possible peripheral nerve hyperexcitability, raising suspicion for Morvan syndrome. Neurological imaging showed diffuse cortical atrophy with chronic small vessel ischemic changes, while serum VGKC panel revealed mildly positive CASPR2 antibodies. A tilt table test confirmed orthostatic hypotension without compensatory tachycardia. The patient was treated with intravenous methylprednisolone and rituximab, leading to clinical improvement with seizure resolution and autonomic stabilization. This case highlights the importance of early recognition and immunosuppressive therapy in CASPR2-spectrum disorders, as well as the need for a multidisciplinary approach in managing autoimmune neurological syndromes.

KEYWORDS: Morvan syndrome, CASPR2 encephalitis, facial-brachial dystonic seizures, autoimmune limbic encephalitis, neuromyotonia, dysautonomia, case report.

CASE REPORT
This case describes a 77-year-old male with a history of coronary artery disease (CAD), systemic hypertension, dyslipidemia, and type 2 diabetes mellitus, who presented with seizures, autonomic dysfunction, and signs suggestive of autoimmune encephalitis. His clinical course included recurrent seizures, orthostatic hypotension, chronic ischemic brain changes, and CASPR2 antibody positivity, raising the possibility of Morvan syndrome, an autoimmune disorder characterized by neuromyotonia, autonomic dysfunction, and central nervous system involvement.[2]

CLINICAL COURSE
First Admission
The patient presented with seizures and loss of consciousness for one week, along with associated nausea. MRI brain and CT cerebral angiogram showed diffuse cortical atrophy with chronic small vessel ischemic changes (Fazekas grade III), along with bilateral AICA vascular loops and compression of the left trigeminal nerve.

Second Admission
The patient was re-admitted for focal seizures involving the right upper limb. MRI brain confirmed previous findings of chronic ischemic changes and vascular abnormalities. EEG was normal, but a tilt table test revealed asymptomatic orthostatic hypotension with no compensatory increase in heart rate, suggesting autonomic dysfunction. Serum VGKC antibody panel showed mildly positive CASPR2 antibodies, supporting the diagnosis of facial-brachial dystonic seizures (FBDS) in the setting of autoimmune limbic encephalitis. He was treated with intravenous methylprednisolone (IVMP, 5 doses), leading to symptomatic improvement. Given the presence of autonomic dysfunction (orthostatic hypotension), central nervous system involvement (limbic encephalitis and seizures), and potential peripheral nerve hyperexcitability (FBDS), a diagnosis of Morvan syndrome was considered.

Third Admission
The patient presented for follow-up with recurrent jerking movements of the right hand and one episode of loss of consciousness one week prior. MRI spine had previously shown spinal epidural lipomatosis at L5-S1, causing severe spinal canal stenosis and cauda equina

compression, contributing to his neurological symptoms. Routine blood tests revealed hyponatremia (128 mEq/L), a common feature of autoimmune syndromes with SIADH-like presentations. He was admitted for rituximab infusion (1g i.v infusion given), after which he remained stable and was discharged. Looking forward for routine follow-up.

DISCUSSION

The constellation of seizures, autonomic dysfunction (orthostatic hypotension), CASPR2 antibody positivity, and possible peripheral nerve hyperexcitability (FBDS) strongly suggests a CASPR2-antibody spectrum disorder, with possible overlap with Morvan syndrome.[7]

- FBDS and Limbic Encephalitis: The patient exhibited FBDS, a hallmark of CASPR2 encephalitis, along with MRI evidence of limbic involvement.[8]
- Autonomic Dysfunction: The tilt table test showing orthostatic hypotension suggests dysautonomia, a key feature of Morvan syndrome.[9]
- Peripheral Nerve Hyperexcitability? The right upper limb focal seizures and persistent jerking movements raise the possibility of neuromyotonia, though further electromyographic (EMG) studies would be needed to confirm.[10]

Given these findings, a working diagnosis of Morvan syndrome overlapping with CASPR2 limbic encephalitis was considered. Early initiation of immunotherapy with IVMP and rituximab led to clinical stabilization.[5]

CONCLUSION

This case highlights a rare presentation of CASPR2-associated autoimmune limbic encephalitis with features suggestive of Morvan syndrome, including FBDS, autonomic dysfunction, and possible peripheral nerve hyperexcitability. The patient's improvement with IVMP and rituximab underscores the importance of early recognition and aggressive immunosuppression in autoimmune neurological disorders [6]. Multidisciplinary follow-up is crucial for ongoing disease management and monitoring for recurrence.

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