Morvan Syndrome Manifesting as Autoimmune Paraneoplastic Encephalitis Associated with Thymoma and Antivoltage Gated Potassium Channel (Leucine Rich, Glioma Inactivated 1) Antibody Detected using F 18 Fluorodeoxyglucose Positron Emission Tomography/Computed Tomography

Abstract
Morvan’s syndrome (MoS) is a rare, complex neurological disorder characterized by neuromyotonia, neuropsychiatric features, dysautonomia, and neuropathic pain. The majority of MoS cases have a paraneoplastic etiology, most commonly thymoma, usually occurring before the diagnosis of the underlying tumor and showing improvement following surgery. We present a case of 60-year-old patient presenting with suspicious of MoS and autoimmune encephalitis (AE), F-18 fluorodeoxyglucose positron emission tomography/computed tomography as single imaging modality detected and confirmed both AE and thymoma.

Keywords: F-18 fluorodeoxyglucose positron emission tomography/computed tomography, Morvan syndrome, paraneoplastic autoimmune encephalitis, thymoma

Introduction
Morvan’s syndrome (MoS), first described in 1890, by the French physician, Augustin Marie Morvan, is a clinical syndrome complex characterized by muscle cramps, loss of weight, autonomic dysfunction, and limbic encephalitis.[1,2] Acquired cases are commonly due to an autoimmune disorder and secondary to the presence of antivoltage-gated potassium channel (VGKC) antibodies.[3] Many patients have an underlying tumor, for example, thymoma, lung cancer, testicular cancer, and lymphoma; this indicates the paraneoplastic nature of the disease.[4] We describe here the interesting image findings of fluorine 18 fluorodeoxyglucose positron emission tomography (PET)/computed tomography (CT) (F-18 fluorodeoxyglucose [FDG] PET/CT) in a unusual case of MoS showing autoimmune encephalitis (AE) pattern in brain PET and thymoma in mediastinum. The patient underwent thymectomy which showed mixed thymoma and he improved in clinical manifestations.

Case Report
A 60-year-old male presented with complaints of tremors, slowness in gait, burning sensation in foot and hand and giddiness. On examination, he had slowness of walking, muscle twitchings all over body, bilateral hand tremors, fluctuations in blood pressure, and decreased urine output. He also had disturbed sleep, irrelevant talk, hallucinations during sleeping, and recent memory loss. With these manifestations of central nervous system, peripheral nervous system, and autonomic dysfunction, MoS was suspected. His anti-VGKC antibody levels (leucine-rich, glioma inactivated 1 [LGI-1] protein) were elevated. Magnetic resonance imaging (MRI) brain was not suggestive of encephalitis. He was referred for whole-body F-18 FDG PET/CT for ruling out paraneoplastic AE. Separate FDG brain PET images [Figure 1] (Scenium software) showed hypermetabolism in bilateral basal ganglia, hippocampus, amygdala, and mesial temporal lobe which is consistent with AE. Whole-body...
images showed mildly hypermetabolic homogenously enhancing lesion in anterior mediastinum measuring 4.5 cm × 3 cm (with a SUVmax of 4) [Figure 2]. He underwent total thymectomy and pathology confirmed mixed thymoma (modified Masaoka Staging IIB).

**Discussion**

MoS is a rare, complex neurological disorder characterized by neuromyotonia, neuropsychiatric features (insomnia, confusion, amnesia, and hallucinations), dysautonomia (hyperhidrosis, severe constipation, drooling, and cardiac arrhythmias), and neuropathic pain. It was first described in 1890 by the French physician Augustin Marie Morvan as “chorée fibrillaire.”[1] Our patient also presented with neuromyotonia, hallucinations, amnesia, and neuropathic pain. Anti-VGKC-complex antibodies are present in the serum of the vast majority of MoS patients, suggesting an autoimmune etiology.[2] Although these antibodies are directed against LGI-1 protein, contactin-associated protein-2 (CASPR-2) or commonly both, anti-CASPR-2 antibodies are predominant and are always associated with thymoma. In fact, patients with MoS may have an associated underlying tumor, such as thymoma (most common), lung cancer, sigmoid cancer, testicular cancer, and lymphoma.[3]

The mechanism through which thymoma triggers autoimmunity has been a matter of debate for several years, and a few different explanations have been proposed. These theories refer to the failure of positive and negative selection of T-lymphocytes in the thymus, resulting in the alteration of the development of T-cells, producing self-reactive lymphocytes.[5] F-18 FDG-PET is a functional imaging modality for in vivo evaluation of the pathophysiology of the brain. It has been reported to reveal abnormal metabolism patterns in AE subjects, such as typical medial temporal lobe hypermetabolism, especially in AE patients with a negative MRI, thus implying that F-18 FDG-PET has higher sensitivity than MRI in the diagnosis of AE subjects.[6] Regional basal ganglia or mesial temporal lobe hypermetabolism on FDG-PET has been observed in LGI-1 AE patients.[7,8] FDG PET/CT is useful to detect primary malignancy in most of the paraneoplastic AE which is most commonly associated with thymoma.[9] Our case presented with typical MoS features and brain PET showed typical AE pattern and detected thymoma which he underwent thymectomy and good improvement in his symptoms.

In conclusion, MoS is a rare paraneoplastic autoimmune disorder characterized by peripheral nerve hyperexcitability, autonomic dysfunction, and sleep disorders. MoS is often a paraneoplastic condition associated with thymoma. Pathophysiologically, MoS is associated with anti-VGKC complex antibodies. F-18 FDG PET/CT is very useful in evaluating AE and also to rule out paraneoplastic etiology as a single modality.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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