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Acute sensory axonal neuropathy (ASAN): An unusual pure sensory variant of Guillain-Barre Syndrome associated with IgM auto antibodies against GM1.

Neurology

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ABSTRACT

Sensory Guillain-Barré syndrome (GBS) is a rare, acute demyelinating neuropathy that presents clinically with involvement of the sensory peripheral nerves only. A 21-year old lady presented with an acute onset of symmetrical sensory abnormalities of all extremities. Neurological examination showed normal motor strength throughout with areflexia, reduced pin-prick and loss of temperature sensation in distal extremities and antalgic gait. A lumbar puncture performed 2 weeks after onset demonstrated albuminocytologic dissociation. Electrophysiology showed sensory neuropathy of axonal type with normal motor nerves studies. Antiganglioside antibodies testing revealed IgM antibodies against GM1. Patient showed dramatic clinical recovery following treatment with intravenous immunoglobin (IVIG).The present case highlights pure sensory variant of GBS with axonal involvement associated with IgM antibodies against GM1.

KEYWORDS

Guillain-Barré Syndrome, IgM antibodies, sensory variant.

Introduction

Pure sensory GBS is a rare variant of GBS that presents clinically with involvement of the sensory peripheral nerve only. The acute, often focal onset suggests an immune-mediated process at the level of the posterior root or dorsal root ganglion. To date, only a few cases of pure sensory GBS have been reported [1, 2, 3, and 4] with the majority of cases exhibiting demyelinating sensory neuropathy. None of the case reports revealed the type of Antiganglioside antibodies demonstrated in the pure sensory GBS. Thus the clinical, electrophysiological and immunological features of pure sensory form of GBS have not been well established, thus resulting in a delay of early diagnosis of these rare variant. Here we report the case of pure sensory form of GBS, with axonal neuropathy, albuminocytologic dissociation and IgM antibodies against GM1 ganglioside with dramatic response to IVIG in a 21 years female.

Case Report

A 21-year female presented with numbness and decreased perception of clothes in distal lower limbs that extended to up to the nipple over a period of 1 week. After a few days she developed symmetrical onset of numbness on both hands and symptoms in lower limbs worsened and she was unable to walk. The patient did not experience any antecedent illness. She had no dry eye or dry mouth symptoms and no bowel or bladder disturbances. Neurological examination showed impaired vibration sensation and dysesthesia below knee up to middle third of leg with areflexia in lower limbs, hyporeflexia of upper limbs and antalgic gait. Sensation in upper limbs, motor strength and rest of neurological examination was normal. Routine blood tests including blood sugar, viral markers, liver function and renal function were normal. Examination of cerebrospinal fluid (CSF) performed 2 weeks after onset showed elevated protein of 188.3mg/dl with normal cells (3 cells with 100% lymphocytes) and normal glucose and chloride levels.CSF findings were consistent with albuminocytologic dissociation. Routine nerve conduction study (NCS) revealed absent sensory potentials in sural nerves with reduced amplitude and normal distal latency in bilateral median and ulnar sensory nerves, while the motor NCS was normal.NCS study was consistent with sensory axonal neuropathy. Magnetic resonance imaging of cervical spine was normal. Vasculitis profile and work up for sjogrens syndrome was negative. In view of involvement of only sensory nerves antiganglioside antibody titre was analysed which revealed IgM antibodies against GM1 ganglioside which further confirmed our diagnosis. Diagnosis of GBS was considered with Brighton level 1 of diagnostic certainty, where patient was unable to walk in view of sensory symptoms .She was given a five day course of IVIG at a dose of 0.4gm/kg/day. Sensory symptoms and gait improved after 10 days of admission. Follow up visit at 3 months showed areflexia in lower limbs with normal gait.

Discussion

The concept of sensory equivalent to ascending paralysis of GBS was first described by Wartenberg in1958 [5].Asbury in 1981 provided diagnostic criteria for sensory loss and areflexia variant of GBS [6]. In 2010 the Brighton Collaboration developed case definitions of GBS with differing levels of diagnostic certainty [7]. The presence of bilateral and relative symmetric flaccid paralysis of limbs was essential for incorporating cases into various levels of diagnostic certainty. Moreover there was no case definition for pure sensory forms of GBS. Therefore it is necessary to reconsider the available diagnostic criteria and add additional criteria for classification of rare regional subtypes of GBS.

Clinical features observed in present case included acute onset symmetrical distal sensory abnormalities, areflexia and antalgic gait with normal motor strength. CSF and electrophysiology was consistent with GBS of axonal type. Acute axonal forms of GBS was first described by Feasby [8], following that an axonal motor variant of GBS termed acute motor axonal neuropathy (AMAN) was reported in 1993 from northern China [9].Reports of acute motor and sensory axonal neuropathy (AMSAN) were published [10] while till date acute sensory axonal neuropathy (ASAN) has not been reported. All the pure sensory forms of GBS described have demyelinating neuropathy [3, 4]. Hence present case highlights acute axonal sensory variant of GBS and we have termed it as ASAN. Presence of IgM antiganglioside antibodies against GM1 has further confirmed diagnosis of GBS and its association with ASAN variant of GBS. Therefore diagnosing sensory GBS and initiating immunotherapy provides early recovery and better outcomes. In conclusion further understanding of the clinical characteristics and the presence of antiganglioside antibodies associated with ASAN need to be studied in future.

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