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CASE REPORT

Guillain Barre Syndrome and polyuria: A rare variant

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Abstract

A 33 year- old gentleman came with a progressive flaccid paraparesis, patchy hyperhidrosis and polyuria following an acute gastro enteritis, ultimately diagnosed as axonal variants-GBS associated with acute pandysautonomia. He was managed successfully with parenteral fluids and desmopressin nasal spray in addition to immunoglobulin.

Background

Guillain Barre Syndrome (GBS) is the most common acute inflammatory peripheral neuropathic disorder. The disease occurs in response to antecedent infections, surgery, immunization and parturition causing antibody mediated neuronal damage mainly to peripheral motor and sensory nerves. The antibodies to GM1, GM1b, GD1a and GalNac-GD1a are in particular implicated in acute motor axonal neuropathy (AMAN). AMAN is commonly reported in young individuals. The incidence of the illness is 1 to 4 cases per 100,000 populations in the US[1]. There are various forms of GBS which manifest as pure motor, pure sensory involvement, autonomic and mixed types. The incidence of pathological subtypes of Acute Motor Axonal Neuropathy (AMAN) and AMSAN neuropathy are virtually alike. Autonomic instability in GBS is not a common presentation which can vary from labile blood pressure to rare bladder or bowel dysfunction.

Key words: Guillain Barre Syndrome, Acute Motor Sensory Axonal Neuropathy, Autonomic instability.

Case report

A previously healthy 33 year old male was admitted to the medical casualty unit with a history of fever and passing loose stools for 4 days. Diarrhea was watery in nature and not associated with abdominal pain. He did not have any respiratory symptoms or urinary symptoms. The patient also complained of body aches. On examination, his pulse rate and blood pressure were 110/ min and 90/70mmHg respectively. He was managed initially with intravenous normal saline. On the fourth day while in the ward the patient complained of bilateral distal lower limb weakness with tingling sensation which was abrupt at onset and rapidly progressive, affected upper limbs as well as respiratory muscles within 24 hours. Neurological examination of lower limbs revealed grade 3/5 power, areflexic limbs with no objective sensory loss. The bedside single breath count assessment revealed 16, 14 and 8 at 0, 4 and 24 hours

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intervals from initial assessment respectively. Cranial nerve examination was normal. There was no significant blood pressure fluctuation or arrhythmias identified during the initial hospital stay and he was fully independent in his bowel or bladder functions. However, three days later, it was observed that he had profuse sweating with erythematous discoloration in the lateral aspect of lower limbs below knees and inner sides of thighs. At the same time it was also observed that his hourly urine output had increased to 250 – 300ml for the next 24 hour. Clinical diagnosis of diabetes insipidus was made by comparing the concentration of sodium in urine and serum. This problem was made under control by desmopressin nasal spray. Initial investigations such as full blood count, ESR, CRP, ALP, AST, ALT, Serum calcium, potassium and sodium were normal. Blood sugar levels were normal and consistent. Lumbar puncture test showed a cellular normal sugar in CSF with elevated protein and the nerve conduction study confirmed AMAN type GBS. The patient was successfully managed with immunoglobulin in addition to mechanical ventilator support for two weeks and regular physiotherapy later. He had achieved almost complete recovery at the second month of illness.

Discussion

GBS is the most common and severe acute paralytic neuropathy. The disease causing severe respiratory paralysis is reported in 30% of cases [2]. Under the umbrella term of GBS several recognizable variants have been identified. AMSAN and AMAN are the two main variant types of GBS. They are characterized by immune attacks directed at axons rather than Schwann cells and myelin. However, the following variations have been reported between them based on clinical presentations, course and prognosis of the disease. AMSAN when compared to AMAN affects more adults than children, is being reported equally affecting in rural and urban communities and having long term functional prognosis with adverse outcome[3].

Dysautonomia in the form of labile hypertension, orthostatic hypotension, sinus tachycardia (most common), sinus arrest, neurogenic pulmonary edema and change in sweat, are well-recognized complications of GBS in 20 % of cases[4]. Quadriplegia and severe proprioceptive sensory loss increase the risk of getting dysautonomia. Besides severe dangerous cardiac complications of GBS neuroendocrine abnormalities are also observed, resulting in polyuria which had been reported in a 16 year - old boy with severe GBS. Polyuria in GBS is multifactorial, partly due to dysregulation of osmoreceptors[5].

The natural history of the illness commences few weeks after a respiratory tract infection or acute gastroenteritis. The pathophysiology of this relatively monophasic illness is identified to be caused by immunological demyelination of nerve fibers by activated lymphocytes and macrophages complex. Symmetrical ascending flaccid paraphrases starting from the distal limb muscle account for the commonest presentation. Several variants of the illness have been described, including Miller-Fisher syndrome with the highest incidence of its presentations like opthalmoplegia, ataxia and areflexia[6]. Other variants of GBS include autonomic dysfunctional types with arrhythmias, fluctuating blood pressures etc.

GBS coupled with polyuria and hyperhidrosis is not commonly reported in variants GBS.

Diagnosis of GBS includes clinical history, additional examination findings and supportive electrophysiological and CSF findings. In most of the instances an antecedent event is noted two to four weeks prior to the illness. This is followed by severe low back pain and progressive asymmetric ascending, selective proximal or distal, weakness along with areflexia which constitute the principal clinical feature of GBS. Nadir of weakness in GBS usually peaks within four weeks of illness. CSF finding of albumin-cellular dissociation is characteristic in GBS.

Overall prognosis of GBS is quite good. However, old age, severe deficit at the onset, cranial involvement, requiring mechanical ventilator, prolonged period of severe paralysis (more than 2 weeks) and electro-diagnostic test of reduced CMAP, fibrillation and polyphasic potentials are identified as bad prognostic factors. Autonomic dysfunction in GBS is considered as one of the major causes of sudden cardiac arrest. Autonomic dysfunction in the form of patchy hyperhidrosis and transient polyuria in GBS are unusual findings.

Management of GBS requires a multidisciplinary team approach in acute phase and neuro rehabilitation stage. Close monitoring of respiratory and cardiovascular parameters are imperative. Plasma exchange or immunoglobulin is proven to play an important role in accelerating the recovery. Corticosteroids have no proven value in GBS.

Take Home Message

GBS is a mono phasic acute progressive sensory- motor polyneuropathy. Severe Guillain Barre syndrome usually causes life threatening autonomic disturbances. Besides dangerous cardiac manifestations, neuroendocrine changes have also been reported with severe GBS, resulting in polyuria. Thus extra attention needs to be paid on close monitoring of fluid balance chart and vital parameters are pivotal to decrease the mortality related to GBS.

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