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Case Report Open Access

Faecal Incontinence in Gullain-Barre Syndrome with Bulbar Palsy-A Case Report with Review of Literature

Sandeep Kumar Kar[1*], Rajat Chowdhury[2], Rajdeep Basu[3] and Soumya Chakrabarti[3]

[1]*Assistant Professor, Cardiac Anaesthesiology, Institute of Postgraduate Medical Education & Research, Kolkata, India*

[2]*Assistant Professor, Anaesthesiology, Institute of Postgraduate Medical Education & Research, Kolkata, India*

[3]*Resident Anaesthesiology, Institute of Postgraduate Medical Education & Research, Kolkata, India*

[*]**Corresponding author:** Sandeep Kumar Kar, Assistant Professor, Cardiac Anaesthesiology, Institute of Postgraduate Medical Education & Research, Kolkata, India, Tel: +919477234900; E-mail: sndpkar@yahoo.co.in

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Abstract

Autonomic dysfunction is common in association with Guillain-Barre Syndrome. Cardiovascular involvement is the most common in this context. But isolated faecal incontinence as parasympathetic hyperactivity is the rarest event. In this case report, we documented a case of Guillain-Barre Syndrome with bulbar palsy and diarrhoea for 3 weeks without detectable pathology and not responding to conventional therapies.

Keywords: Guillain-Barre Syndrome; Acute inflammatory demyelinating polyradiculoneuropathy; Bulbar palsy; Faecal incontinence

Introduction

Guillain-Barre Syndrome (GBS) is explained under common causes of acute poly-neuropathy in adults. Acute Inflammatory Demyelinating Polyradiculoneuropathy is a common variant in Guillain-Barre Syndrome. It is slightly predominant in males than females [1]. The disease generally starts as ascending neuropathy from the lower extremities, and involves upper extremities and respiratory muscles over a short period of time, with a history of upper respiratory or gastrointestinal infections in the 1-4 weeks prior to onset in majority of patients [2]. However in Guillain-Barre syndrome variants, some patients have unusual distribution of muscle involvement [3].

Autonomic dysfunction has been reported in association with Guillain-Barre Syndrome in as many as 66% of patients in a study by Singh NK et al. [4]. Both parasympathetic and sympathetic system may be disturbed. They have shown incidences of sinus tachycardia (33.3%), bradycardia (8.3%), hypertension (33.3%), postural hypotension (35%), urinary sphincter disturbances (20.8%) and anhydrosis of lower limbs (12.5%) of cases in their study.

Here we are going to present a case of Guillain-Barre Syndrome with bulbar palsy associated with faecal incontinence. Our case is different in that in addition to having bulbar palsy which is rare our patient also had faecal incontinence as autonomic dysfunction which is further rarer.

The Case

A 36 years old non-diabetic, non-hypertensive male patient presented with sudden onset weakness in all limbs associated with drooling of saliva, difficulty in swallowing and speech after 15 days of low grade, intermittent fever with mild shortness of breath. His past medical and surgical history were insignificant. There was no similar history of limb weakness from his family background. His immunization was completed and no recent history of vaccination. He had no history of chronic drug intake.

The nadir attained within 3 weeks of the starting of the weakness. The patient developed progressive thinning of limb muscles and increased difficulty in breathing. He was transferred to Intensive Therapy Unit. The patient was intubated and kept in assist controlled mode of ventilator support.

During the 3rd week of this event, the patient developed excessive passage of loose stool.

On general examination, the patient was alert and GCS- $E_4V_5M_6$, heart rate: 90 bpm, regular, blood pressure: 126/78 mm Hg. Neurological examination was suggestive of bilateral symmetrical flaccid quadriparesis, tone and power of all four limbs were decreased, deep tendon reflexes were all absent. Cranial nerve examination revealed bulbar palsy in the forms of dysphagia, dysphonia, nasal regurgitation. Pupils were bilaterally tonic, light reflex was absent, but there were no ophthalmoplagia, ptosis or intra-ocular pathology. There was no sensory deficit and spine and cranium were normal. Bladder functions could not be elicited as the patient was catheterized. Per rectal examination showed decreased sphincter tone.

Complete hemogram and serum electrolytes level were within normality. On day 10 of this event Cerebrospinal fluid study showed protein: 128 mg/dl, mononuclear cells: 8 /cu mm, glucose: 93 mg/dl (plasma glucose level was 98 mg/dl). Serial Cerebrospinal fluid analysis was reported as albuminocytological dissociation. On the same day, Magnetic Resonance Imaging brain shows no abnormality.

On day 12, electro-physiological study showed bilateral decreased in distal latency, conduction velocity with absent H reflex and F wave. Normal CAMP and SNAP sparing sural nerve.

After diagnosing the case I.V. immunoglobulin was started as 400 mg/kg body weight/day for consecutive 5 days. Besides this cardiac monitoring, nutritional support, Deep Vein Thrombosis prophylaxis, postural care, oral care, physiotherapy, speech therapy was undertaken.

Event of diarrhoea was associated with painless passage of loose flakes of stool without any blood tinge. Stool was sent for routine examinations for detection of ova, viral antigen detection, culture and Clostridium difficile toxin A and B assay. Bowel care had been undertaken as dietary changes (less fat, less whole grain, less lactate, high fibre), antibiotic coverage (for aerobic, anaerobic bacteria and parasites), pre-biotic and pro-biotic supplements, anti-secretary agents. Serum electrolytes were monitored and supplemented accordingly. No pathogen was found on stool analysis to be offended, routine examination was also within normal range. Colonoscopy was done to rule out non-infective local pathology but no abnormality was detected. Diarrhoea did not respond to supportive therapy and persisted for 3 weeks but it was getting resolved spontaneously as the patient was regaining power and tone of all limbs and respiratory muscles as well.

Spontaneous breathing trials were continued with close vigilance of respiratory rates, respiratory pattern, involvement of accessory muscles, tidal volume, vital capacity, breath holding time, negative inspiratory force and spirometry module based monitoring of FEV_1. The patient was successfully weaned off from ventilator after 4 weeks without any kind of complication and sent back to general ward.

In the ward, rehabilitation was continued in forms of cough exercise, speech therapy, proprioceptive neuromuscular facilitation to improve muscle power, joint movements and was trained to achieve normal phonation and deglutition. Patient was discharged and shifted to local centre for above mentioned rehabilitative care.

Discussion

This case report highlights the rare association of faecal incontinence with Guillaine-Barre Syndrome and we shall like to substantiate the findings that we gathered from the case scenario.

Postulated immunopathogenesis of Guillain Barre Syndrome: [5]

B cells recognize glycoconjugates on the offender (e.g. C. Jejuni) that cross-react with ganglioside present on Schwann cell surface and subjacent peripheral nerve myelin. Some B cells, activated via a T cell–independent mechanism, secrete primarily IgM . Other B cells are activated via a partially T cell–dependent route and secrete primarily IgG; T cell help is provided by CD4 cells activated locally by fragments of Cj proteins that are presented on the surface of antigen-presenting cells (APC). A critical event in the development of Guillain-Barre Syndrome is the escape of activated B cells from Peyer's patches into regional lymph nodes. Activated T cells probably also function to assist in opening of the blood-nerve barrier, facilitating penetration of pathogenic autoantibodies. The earliest changes in myelin (right) consist of edema between myelin lamellae and vesicular disruption of the outermost myelin layers. These effects are associated with activation of the C5b-C9 membrane attack complex and probably mediated by calcium entry; it is possible that the macrophage cytokine tumor necrosis factor (TNF) also participates in myelin damage.

In case of Axonal injury there is first attack at motor nodes of Ranvier; macrophage activation, few lymphocytes, frequent periaxonal macrophage but the extent of axonal damage is highly variable and predominantly mediated by anti-GD1a antibodies [5].

Considering Brighton case definition for Guillain-Barre Syndrome, this case meets level 1 diagnostic certainty in all components of criteria in view of history, clinical examinations, Cerebrospinal fluid analysis and electro-physiological study [6].

In this case the atypical findings are bulbar palsy and faecal incontinence. Bulbar palsy may be associated with several other neurological, even some non-neurological disorders. Among them botulism, diphtheria, myasthenia gravis, multiple sclerosis, brainstem vascular abnormalities, tumour invasion are important, even in Pharyngeal-cervical-brachial (PCB) variant of Guillain-Barre syndrome [3].

Pharyngeal-cervical-brachial (PCB) variant of Guillain-Barre syndrome is presented with oropharyngeal, neck, and upper limb muscle involvement [3]. The incidence of pharyngeal-cervical-brachial variant is also not much common [3]. This variant does not cause generalized limb weakness also it is commonly associated with axonal variety rather than demyelinating polyradiculoneuropathy [7]. Raised CSF proteins and demyelinating features on nerve conduction studies are the other features to support the diagnosis of the case.

Case studies have shown that association of early bulbar palsy with Guillain-Barre Syndrome and preceding history of respiratory symptoms in Anti GM-3 and Anti GT-1b positive patients and Anti GT-1a positive patients [8,9]. But this test is much expensive and not available in our hospital, so it could not be done.

Now in the question of faecal incontinence, the authors excluded all common infective causes like bacterial, viral, parasites by stool culture, detection of ova, viral antigen study. Diarrhoea was not responding to conservative treatment. Also there was no evidence or history of drug used for Cl. difficile related diarrhoea. No local pathology was detected in colonoscopy. Faecal incontinence was resolving with improvement from weakness and was fully cured on gaining muscle power. So, taking all the points under consideration, it can be hypothesised that this event was due to parasympathetic hyperactivity [10] on lower gut as autonomic involvement in association with Guillain-Barre Syndrome.

Dysautonomia is common association in Guillain-Barre Syndrome [11], involving both parasympathetic and sympathetic nervous system. Among autonomic involvements it affects most commonly cardiovascular system [12], presents as sinus tachycardia, labile hypertension and postural hypotension. However, sinus bradycardia, asystole, supraventricular tachycardia, junctional tachycardia and ventricular tachycardia have also been reported in studies [13], even serious bradyarrhythmias in less severely affected patients [14]. Parasympathetic hyperactivity is usually intermittent, causing fatal bradyarrhythmias ranging from bradycardia to asystole, and a significant number of deaths in Guillain-Barre Syndrome patients [11].

Also Guillain-Barre Syndrome may involve G.I. autonomic nervous system causing gastro-paresis [11] and adynamic ileus (17 out of 114 patients) [15]. Autonomic dysfunction may also be manifested as SIADH (Syndrome of Inappropriate Anti Diuresis) [16,17].

Manifestations of autonomic involvements differ qualitatively between Acute Inflammatory Demyelinating Polyneuropathy and Acute Motor Axonal Neuropathy. The former one is characterized by cardio-sympathetic over-activity, excessive or reduced sudomotor function and preserved skin vasomotor function, while Acute Motor Axonal Neuropathy is not generally associated with such marked autonomic dysfunction, only decreased sudomotor function in patients with severe neurological derangement [18].

The occurrences of dysautonomia increase with quadriplegia, respiratory failure or bulbar involvement in Guillain-Barre Syndrome [19]. Though mechanism is not clearly understood, pathology of the Gastrointestinal autonomic dysfunction in Guillain-Barre Syndrome usually reveals immune-mediated inflammation in autonomic ganglia with destruction of peripheral ganglionic cells [15] causing imbalance between sympathetic and parasympathetic activity. Demonstration of mononuclear cell infiltration, demyelination, chromatolysis, and nodules of Nageotte within autonomic ganglia in histopathology [11] further strengthen the hypothesis.

Thus, this case is unique in the sense of isolated faecal incontinence for autonomic dysfunction in Guillain-Barre Syndrome with bulbar palsy and to our knowledge this is the first case reported in medical literature with such atypical pictures of dysautonomia.

Conclusions

From the discussion above, the authors conclude that though rare, isolated faecal incontinence may be present with Guillain-Barre Syndrome as autonomic dysfunction. More studies are necessary in this field to understand the true pathophysiology of such presentation. Supportive care should be continued till full recovery of cases.

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