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Unusual Variant of Guillain-Barré Syndrome Following Hepato-biliary Surgery — A Rare Case Report

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Abstract

Background: Guillain-barré syndrome (G.B. Syndrome) is an acute inflammatory poly-radiculoneuropathy characterized by weakness and areflexia typically following viral infection, vaccination, and rarely surgery. Acute Inflammatory Demyelinating Poly-radiculoneuropathy is the most common subtype of G.B. Syndrome. Although post-operative G.B. syndrome is a rare entity, there are few case reports of G.B. syndrome after gastric surgery. But there have been no reported case scenarios of atypical variety of this neurologic entity following hepato billiary surgery. Hence our objective is to put forward this message to the readers.

Case Summary: Here we are going to report an unusual case of G.B. syndrome in a 45 years old female, identified in early post-operative period following Hepato-biliary surgery. Patient was diagnosed as AIDP with facial and occulomotor nerve palsies. She needed ventilator support for 6 weeks and weaning from ventilator was successful.

Rare variant of G.B. Syndrome puts clinicians in a dilemma that too when happens in a post-operative scenario. Through this case report authors aim at close vigilance over the patients during the post-operative periods with sound clinical judgment to detect such rare incidence at the earliest and intervene without delay.

Keywords: Hepato-biliary surgery; General anesthesia; AIDP; Facial and Occulomotor nerve palsies; Ventilator

Introduction / Background

In the developed countries the median incidence of Guillaine Barre syndrome has been estimated to be 1.11 per lakh population. The male to female gender ratio has been reported as 1.78:1. It is an acute, frequently severe & fulminant poly-radiculoneuropathy seeking urgent intervention. It is associated mostly following respiratory tract infection (40%), gastrointestinal infection (20%) [1]. Weakness may involve respiratory muscles and patients may have to be managed with ventilator support in critical care set up. Death has been documented in 4-15% patients with Guillaine Barre Syndrome [2] and 12-20% of patients with GBS may require ventilator support for respiratory paralysis [3]. Within those ventilated patients, 20% [4] may die due to Ventilator associated Pneumonia, ARDS, Sepsis.

Post-surgical neuropathies are usually attributed to mechanical factors, such as compression, stretch, contusion or transection. Surgery is documented as a rare cause of GBS and there are case reports of Guillaine Barre Syndrome occurring on Days 4, 7, 9, 11 [5-7] and even after 2 weeks [8] following surgeries. Also in the medical literature some cases are reported as Guillaine Barre Syndrome following surgeries done under epidural anesthesia [9-11]. No case report of such early onset GBS has been documented so far after Hepato-biliary surgery under general anesthesia.

Here we documented a rare case of unusual variant of G.B. syndrome including cranial nerves involvement from 3rd postoperative period following a Hepato-biliary surgery done under GA.

The Case

A non-diabetic, non-hypertensive 45 years old female patient presented with sudden onset weakness of all four limbs accompanied with drooping of left upper eyelid, facial weakness and deviated tongue from 3rd postoperative day following Hepato-biliary surgery for Extra-hepatic biliary stricture and nadir attained within 2weeks. She underwent General anesthesia using Injection Thiopent-

one (250mg) as induction agent and Inj. Atracurium (0.5mg/kg—loading and 0.2mg/kg/hour—maintenance dose) as muscle relaxant. Maintenance of anesthesia done by N_2O:O_2 (5:3) and inhalational agent Isoflurane 0.2% MAC. There was neither history of recent respiratory tract infection nor gastrointestinal infection. She had a previous history of Cholecystectomy and Choledocho-duodenostomy 4 years ago for CBD stone which was uneventful.

There was no history of such neurological disorder running in the family. The patient was not from any endemic area of viral diseases. Her immunization status was completed and no recent history of vaccination was there.

Progressively patient developed both sided lower limbs muscle thinning and shortness of breath. Patient was shifted to ITU on 5th postoperative day and kept in ventilator at assist controlled mode.

Neurological examination revealed her to be alert with normal higher functions and bilateral symmetrical ascending flaccid quadriparesis. Tone and power of the both lower and upper limbs were diminished ($power_{upper\ limbs}$-2/5, $power_{lower\ limbs}$-1/5). All deep tendon reflexes were absent. Bilateral planter responses were nonreactive. Cranial nerve examinations showed drooping of left upper eye lid (Ptosis—left occulomotor palsy) and bilateral asymmetric lower motor type of facial nerve palsy. Also there was apparent deviation of tongue to the left without atrophy Pupils are bilaterally tonic and not reacting to lights. Sensory functions were normal. There was no abnormality with spine and cranium. Bowel and bladder disturbances could not be evaluated.

There was no evidence of sepsis in routine blood investigation. Hb-10gm%, TLC-7200/cu mm (N^{84} L^{12} E^{03} M^{01} B^{00}) Platelet- 1, 65,000/cu mm. Blood, sputum and urine culture showed no growth of organism. Serial monitoring of serum electrolytes had been done and reports were within normal range. Excised materials after surgery had been sent for histopathological examination and no malignancy was detected.

CSF study revealed –Protein- 148mg/dL, Mononuclear cells - 6/cu mm, Glucose- 70mg/dL with Plasma glucose 95mg/dL on Day 5 after onset of symptoms. Serial CSF studies showed increasing trends of protein with few mononuclear cells.

On Day 6 after onset of symptoms, Motor nerve conduction study showed normal CAMP amplitude, distal latency and conduction velocity in both Common peroneal nerves, Tibial nerves, Median nerves and Ulnar nerves. 'F-wave' study revealed absent F-wave in both Median, Ulnar and Sural nerves. Sensory nerve conduction study showed normal Sural SNAP amplitude and absent SNAP amplitude in both Median and Ulnar nerves. 'H reflex' study showed absent H-wave bilaterally. Repetitive Nerve Stimulation Test showed no significant decremental response in any of the studied nerves. So the Nerve conduction velocity report suggestive of Sural nerve sparing AIDP.

Immediately after diagnosis treatment with I.V. immunoglobulin was initiated and given as total dose of 400mg/kg body weight/ day for 5 days. Simultaneously respiratory therapy, cardiac monitoring, physiotherapy, nutritional supplementation, postural care, DVT prophylaxis were given. Physiotherapy was given as gentle passive movements through full ROM at least three times a day especially at hip, shoulder, wrist, ankle, feet, effleurage massage to lower limbs, avoidance of prolonged hip and knee flexion for maintenance of normal joint movements and blood circulation; postural drain areas of lung tissues, 2-hourly turning into supine or side lying positions, manual techniques like vibration with/ without over pressure, rib springing to stimulate cough and to maintain airway clear. Also 2 hourly position changes had been undertaken to prevent pressure sore.

With treatment, power and tone of both upper limb and lower limb were improving. Weaning trials from ventilator were carried on with continuous Spirometry module based monitoring of FEV_1, Negative inspiratory force, Respiratory rate, Vital Capacity, Tidal volume, Breath holding time and she was taken out of ventilator successfully with spontaneous normal breathing pattern and without complication after 6 weeks of ventilator support. The Patient was shifted to the ward after gaining normal neurological function. Patient was taught effective cough exercise and proprioceptive neuromuscular facilitation technique was under taken to improve muscle power and joint movements after weaning from ventilator support. Patient was discharged and transferred to rehabilitation centre.

30 days later review by our critical care and neuro rehabilitation team suggested that her recovery was satisfactory and she can escalate her exercising capabilities gradually. Gastro enterology surgical team described this as their first endeavour.

Discussion

Guillaine Barre Syndrome is the one of the most common causes of acute poly-radiculoneuropathy in adults. It can occur at any age with slight predominance in males [12]. The disease begins in the lower extremities, and over the course of hours or days, it ascends, characterized by weakness in the arm and facial muscles. The majority of patients have a history of upper respiratory tract or gastrointestinal system infections in the 1-4weeks prior to symptoms [13]. While the pathogenesis of GBS is unknown, it is accepted as a hypersensitive humoral and cellular immune response, generally attacking peripheral nerve system components [12]. Recently, the occurrence of GBS after major and minor surgical operations has been increasingly debated. The relevant literature is limited to case reports [14].

So, from the case discussed above, the following differential diagnoses should be taken under consideration—
1) Critical illness poly-neuropathy
2) Myasthenia gravis
3) Dyselectrolytemia

We can exclude Critical illness poly-neuropathy (CIP) because this patient acutely presented limb and respiratory muscle weakness that too with minimal period of immobilization. CIP is a complication of sepsis and multi-organ failure in 70% patients presenting with difficulty in weaning from ventilator and varying degree of limb weakness [15]. We believe that this patient had G.B syndrome distinct from CIP or other myopathies for several reasons –Firstly, there was no evidence regarding sepsis or systemic inflammatory response syndrome during limb weakness on postoperative days. There was no evidence of septic encephalopathy as well and her higher functions were normal. Secondly, there was no evidence of any bacterial, viral or fungal infection. Thirdly, CSF study has shown typical albuminocytological dissociation. Electrophysiological study revealed demyelinating poly-radiculoneuropathy sparing sural nerve. The absence of F-wave in nerves with normal CMAP amplitude is highly specific for demyelination [16]. The case was atypical in asymmetric involvement of 7^{th} and 3^{rd} cranial nerves.

Dyselectrolytemia also cannot be considered, it is excluded on the basis of patient's higher functions and normal serum electrolytes level during serial monitoring. There was no history of prolonged use of steroid which may cause the same clinical picture [17].

Anesthetic agents are often responsible for prolonged muscle weakness but here general anesthesia was given in the forms of Thiopentone, Atracurium, N_2O and Isoflurane and none of these is documented for such weakness.

AIDP may be caused by primary neoplasms and lymphomas [18] but here pre-operative clinical presentation, per operative surgical access and post-operative biopsy findings showed no evidence of malignancy.

Occulomotor involvement hints towards Myasthenia gravis with almost same clinical features but it can be excluded on basis of typical CSF finding & EMG-NCV reports suggestive of AIDP. Also pupillary involvement is suggestive of AIDP as cilliary muscle involvement does not usually occur in Myasthenia. Moreover there are some case reports suggesting ptosis in association of AIDP in a clinical study in Taiwan [19]. But exact pathophysiology of this atypical presentation is unknown.

The question of bulbar palsy may arise due to deviation of tongue but it was apparent due to asymmetrical facial weakness and without atrophy of tongue.

The pathophysiology of postoperative GBS has not been clear yet. The traumatic event presumably triggers the immune response. Histology reveals macrophage infiltration leading to inflammation into the nerve [20]. Two immune response processes have been proposed to explain the infiltration process. One is T cell response against antigen on nerve surface causing inflammatory mediator release [21]. Another is humoral response from antibodies binding to epitopes on the nerve surface by complement activation [22]. Though there is no clear evidence yet, it can be hypothesized that inflammation can be induced by ischemia, general trauma after surgery which consequently cause humoral & cytokines response by the immune system [23] and lead to GBS. These may explain the variable duration of onset after surgery and disease course.

On the setting of postsurgical AIDP, 6 cases among 93 in a study group had been found to have a surgery within 6 weeks prior to GBS [24] and they calculated the Relative risk of G.B Syndrome within 6 weeks of surgery as 13.1 times greater than normal incidence in study population. In the medical literature, surgeries associated with GBS so far include cardiac surgeries [5,25], spinal surgeries [6,26], hip arthroplasty [8], cranial surgeries [27], oral surgeries [28], even gastrectomy [7], with various durations of onset of symptoms.

But the peculiarity of this patient enlightens us that such case scenarios need to be documented further and emphasizes that even minimal trauma surgeries can trigger the inflammation system so as to precipitate Acute inflammatory demyelinating polyneuropathy.

Conclusion

Though respiratory and gastrointestinal infections have a major potentiality to cause AIDP, Hepato-biliary surgeries provoking such altered immune status induced polyradiculoneuropathy needs to be documented in a larger scale with more researches. Such case scenarios further strengthen the fact that acute onset neurologic insults are increasing post operatively too and skilled anaesthesiologists in post-operative rounds can detect them early to help patient recovery. Atypical presentation of Guillaine Barre Syndrome though documented but pathophysiology needs to be uncovered.

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