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-radiculo-neuropathy characterized by weakness and areflexia typically following viral infection, vaccination, and rarely surgery. Acute Inflammatory Demyelinating Poly-radiculo-neuropathy 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 neurolologic entity following hepato biliary 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-radiculo-neuropathy 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-
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 1-4 weeks 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 septice 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-radiculooneuropathy 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 7th and 3rd 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, N₂O 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.

Occlusomotor 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 ciliary 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.

**References**