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Anesthetic Challenges in Hirayama Disease Patients Undergoing Cervical Spine Surgery—A Case Series

Sreyashi Naskar[1] Soumya Chakrabarti[1] Dipanjan Dawn[1] Amita A. Pahari[1]

[1] Department of Neuroanaesthesiology, Bangur Institute of Neurosciences, Kolkata, West Bengal, India

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Address for correspondence Sreyashi Naskar, MD, Department of Anaesthesiology, Bangur Institute of Neurosciences, 52/1A Sambhunath Pandit Street, Bhowanipore, Kolkata 700020, West Bengal, India (e-mail: sreyashi.n@gmail.com).

Abstract

Hirayama disease (HD) is a rare disease, resulting from cervical compressive myelopathy, manifesting as upper limb muscular atrophy, and rarely autonomic and upper motor neuron signs. Anesthesia management is challenging—careful neck positioning during bag-mask ventilation and endotracheal intubation, avoidance of drugs that release histamine, multimodal monitoring to avoid delayed recovery, anticipation of hypotension, and blood loss due to autonomic dysfunction—all this is necessary for successful outcome of general anesthesia in HD patients. This case series demonstrates that preexisting autonomic dysfunction in HD patients should alert the anesthesiologists regarding higher likelihood of hemodynamic perturbations and blood loss, compared with patients who have normal autonomic functions, and henceforth take appropriate precautionary measures.

Keywords
- Hirayama disease
- motor neuron disease
- spinal
- muscular atrophy
- nonprogressive

Introduction

Hirayama disease (HD) is a rare[1] neurological condition characterized by insidious onset of uni- or bilateral muscular atrophy and lower motor neuron (LMN) weakness of predominantly the distal upper extremities, typically affecting adolescent Asian males.[2] Sensory, autonomic system involvement and upper motor neuron (UMN) signs are rarely seen.[3] Dr. Hirayama first reported the disease in 1959, and described the pathophysiology of the disease, which includes loss of the normal cervical lordosis, flattening of the spinal cord in the neutral position, and anterior displacement of the cord during flexion, accompanied by increase in intramedullary pressure due to the dorsal epidural fat proliferation and venous plexus engorgement. These features cause dynamic compression of the cervical cord, resulting in recurring transient ischemic damage to the anterior horn cells (AHCs).[4] There remains a paucity of studies describing the unique anesthetic challenges in HD in the background of distinct physiological aberrations in their neuromuscular, cardiovascular systems and airway-related difficulties.

We share our experience with three HD patients undergoing cervical spine surgery under general anesthesia, detailing individual clinical presentations, perioperative challenges, and management strategies. We have elaborated on the vigilant care required specifically in HD patients with autonomic dysfunction under general anesthesia.

Case Report

Three adolescent male patients with HD visited the neuromedicine clinic with varying grades of upper limb LMN signs, complaining of progressive weakness of their hand muscles, leading to difficulty in performing daily activities. After a thorough clinical examination and neurological workup, they were subjected to conservative management (cervical collar) for varying periods ranging from 1.5 to 2 years, but it failed to arrest symptom progression. Subsequently, they

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Table 1 Preoperative findings

Demographic and other characteristics		Case 1	Case 2	Case 3
Age		18 y	21 y	21 y
Gender		Male		
Profession		Laborer	Student	Student
Symptoms		1. Weakness of left hand and forearm, gradually progressing from distal to proximal	1. Progressive weakness and thinning of right forearm and hand 2. Difficulty in fine movements of right hand, like writing	1. Progressive weakness and thinning of both (right > left) forearms and hands 2. Difficulty in fine movements of right hand, like writing 3. Difficulty in lifting heavy objects
Duration of symptoms		3 y	1.5 y	2 y
Signs	Power	4/5 over his left hand and forearm muscles, with thenar and hypothenar atrophy	4/5 over his right hand and forearm muscles, with thenar and hypothenar atrophy	Right side—3/5, left side—4/5 (over his hand and forearm muscles), with thenar and hypothenar atrophy (right > left) (►**Fig. 1A**)
	Skin temperature	Cold extremities	–	–
	Spasticity	–	–	Present in right upper limb
	Bedside test of autonomic function heart rate and blood pressure responses to standing	(30:15) ratio < 1.04 and supine SBP—standing SBP >10 mm Hg	(30:15) ratio > 1.04 and supine SBP—standing SBP < 10 mm Hg (normal findings)	(30:15) ratio < 1.04 and supine SBP—standing SBP >10 mm Hg
	Deep tendon reflexes	Normal		
Airway	Modified Mallampati grade	II		
	Neck movements	Adequate		
	Loose or buck tooth	None		
Investigations	Complete hemogram	Normal		
	Serum electrolytes			
	Coagulation profile			
	Chest X-ray			
	Electrocardiogram			
Dynamic MRI of spine	Findings	Anterior displacement of the posterior dural wall during flexion and spinal cord compression along with a crescent-shaped enhancement of the posterior epidural space (Crescent moon sign, ►**Fig. 1B**)		
	Levels of spinal cord compression	C3–C7	C3–C7	C4–C7
NCS/EMG		Signs of chronic motor axon denervation with likely site being the AHC	Decreased amplitude of the compound muscle action potential of right ulnar and median nerves	NCS-decreased amplitude of the compound muscle action potential of bilateral ulnar and median nerves Needle EMG of the forearm and hand muscles were suggestive of bilateral C7–T1 radiculopathy and focal neuropathy

Abbreviations: AHC, anterior horn cell; EMG, electromyography; MRI, magnetic resonance imaging; NCS, nerve conduction study; SBP, systolic blood pressure.

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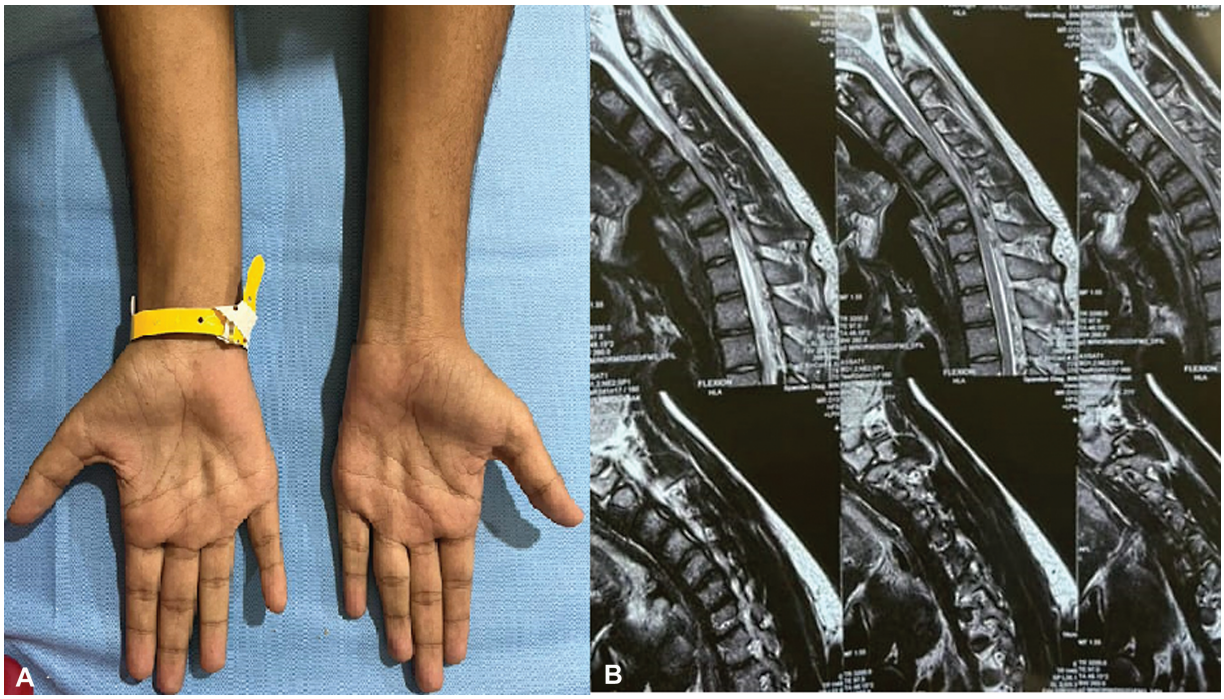

Fig. 1 (**A**) Wasting of thenar and hypothenar muscles of third patient (right > left), (**B**) T2-weighted sagittal magnetic resonance image of spine of first patient, in flexion shows anterior displacement of the dorsal dura compressing the thecal sac, with a prominent dorsal epidural compartment.

were planned for cervical lateral mass fixation and duraplasty surgery under general anesthesia.

Preoperative evaluation showed these findings (►**Table 1**).

Inside the operating room, standard American Society of Anesthesiologists monitoring along with bispectral index (BIS), train of four (TOF), and invasive arterial blood pressure monitoring, to guide fluid and vasopressor therapy, were initiated. Two wide bore peripheral intravenous access were established. The plan of airway management was King Vision channeled video laryngoscopic intubation with flexometallic endotracheal tube under general anesthesia. A difficult airway cart including appropriate-sized fiberoptic bronchoscope was kept ready. A neutral head position was ensured during bag-mask ventilation as well as laryngoscopy. Preloading with 5 mL/kg crystalloid was followed by premedication with glycopyrrolate 200 µg. Anesthetic induction was done using fentanyl (2 µg/kg), propofol titrated to loss of verbal response, and rocuronium (1 mg/kg), after confirmation of adequate bag-mask ventilation and a target BIS of 50. Video laryngoscopic intubation was done. After padding of eyes and pressure points, the patients were made prone using manual in-line stabilization maneuver and placed in neutral head position. Autocompression stockings were applied on the lower limbs for deep venous thrombosis prophylaxis. Depth of anesthesia was maintained with desflurane, targeting BIS of 40 to 60. TOF-guided boluses of rocuronium were given. When surgery was over, patients were kept in supine position, and TOF-guided reversal of neuromuscular blockade was done and extubated with cervical collar in situ.

The patients were subject to regular physiotherapy and discharged on postoperative day 5. During discharge, the neurological status of the patients was similar to preoperative period; however, 6 months later, all three patients had significant improvement in grip strength, and the third patient had increased motor power (3/5 to 4/5) in his right hand.

Discussion

HD differs from classical types of motor neuron disease[5] because of its nonprogressive course, selective involvement of upper limbs, and pathologic finding of chronic microcirculatory changes in the territory of anterior spinal artery supplying the AHCs of lower cervical cord.[6] The disease poses a diagnostic challenge due to its rare incidence and overlap with related disorders. Apart from Tashiro et al[2]'s diagnostic criteria, another modality is the typical "crescent moon sign" seen in dynamic flexion magnetic resonance imaging.[7]

All our patients satisfied majority of Tashiro et al's criteria; however, the first and third patients had autonomic dysfunction. Third patient additionally showed right upper limb spasticity. Autonomic dysfunction and UMN lesions are uncommon clinical manifestations of HD.[8] Of note, is the occurrence of greater degree of hemodynamic fluctuations and significant blood loss in patients with preexisting autonomic dysfunction, in the present case series (►**Fig. 2A**). Both the first and third patients had more hypotension episodes and greater blood loss (first patient required blood transfusion), compared with the second patient. Since parasympathetic dysfunction is associated with gastroparesis and reflux, antacid and prokinetic maybe considered as premedications.[9]

Since pathophysiological basis of HD is cervical flexion myelopathy,[4] neutral neck position is preferred for airway

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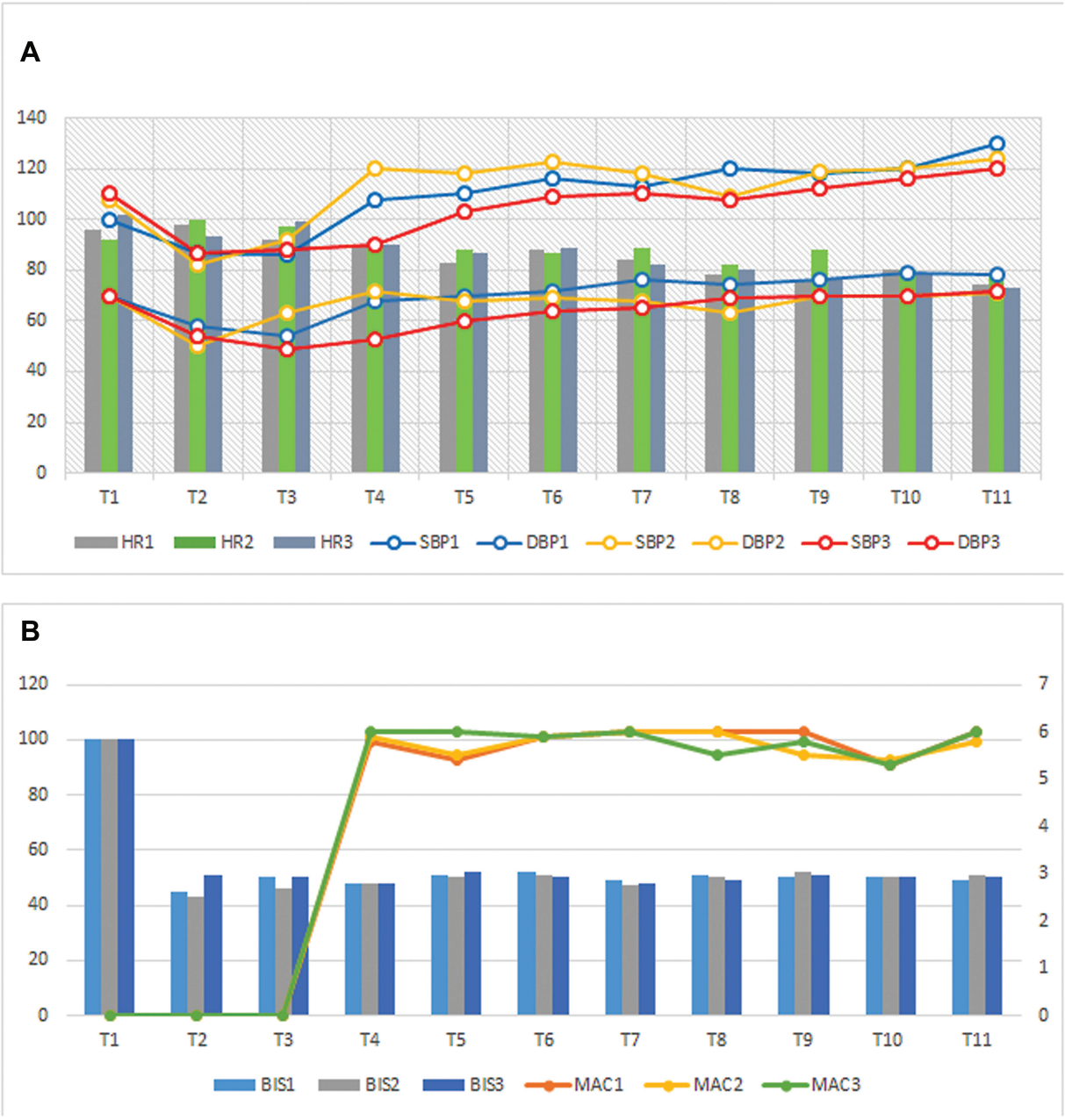

Fig. 2 (A) Blood pressures and heart rates of the three patients at different points of time. (B) BIS and MAC of the three patients at different points of time. BIS, bispectral index; MAC, minimum alveolar concentration (the numbers 1, 2, and 3 correspond to first, second, and third patients, respectively); SBP, systolic blood pressure; DBP, Diastolic blood pressure; HR, heart rate; T1, baseline, before induction; T2, immediate postinduction; T3, immediately after proning; T4, 15 minutes after T3; T5, 15 minutes after T4; T6, 15 minutes after T5; T7, 15 minutes after T6; T8, 15 minutes after T7; T9, 15 minutes after T8; T10, 15 minutes after T9; T11, 15 minutes after T10.

management and surgery. Cervical collar was in place during intubation, positioning, and extubation. Careful padding of pressure points before proning due to coexistent muscle atrophy and spasticity is important.

These patients may have reduced physiological reserve to tolerate hypotension-induced ischemia and require higher arterial pressures to maintain spinal autoregulation.[1] Hence, adequate preloading was done and normovolemia ensured by goal-directed (EV 1000 monitor) fluid therapy targeting a mean arterial pressure above 90 mm Hg. Hypotension was treated by direct-acting α-adrenergic agents (phenylephrine 0.15–1.5 µg/kg/min infusion) in the background of autonomic dysfunction. Forced air warmers have been used, as these patients are prone to hypothermia. Many of them have coexistent airway allergies with eosinophilia.[10] Therefore, administration of histamine-releasing drugs should be avoided. In patients with denervation because of spinal cord lesions, increased number of perijunctional nicotinic acetylcholine receptors on skeletal muscles can cause hyperkalemia after succinylcholine, while long-acting muscle relaxants are associated with a prolonged neuromuscular blockade. Judicious use of

anesthetic agents, guided by BIS (►**Fig. 2B**) and neuromuscular monitoring, is essential to prevent delayed recovery. In the present report, diseased ulnar and median nerve necessitated TOF monitoring in the sural (posterior tibial) nerve and flexor hallucis muscle. Postoperatively, adequate analgesia, antiemesis, and physiotherapy should be considered.

Conclusion

HD presents unique challenges for the anesthesiologist, with respect to airway, autonomic system, cardiovascular system. A vigilant hemodynamic management is warranted in cases with preexisting autonomic dysfunction. Meticulous care is crucial in manipulation and securing of the airway, positioning, maintenance of stable hemodynamics, and successful extubation during cervical spine surgery.

Conflict of Interest
None declared.

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