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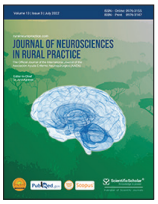

Case Report

Anesthetic challenges in a patient with Hirayama disease with quadriparesis and autonomic dysfunction undergoing cervical spine surgery

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ABSTRACT

Hirayama disease is a rare neurological disorder, characterized by muscular atrophy of the distal upper extremities. The occurrence of spastic quadriparesis and autonomic dysfunction is rarely reported and has important perioperative considerations during cervical spine surgery for the treatment of this disorder. The role of the anesthesiologist is vital in the thorough assessment of the patient for the involvement of the pyramidal tract, autonomic dysfunction, gastroparesis, hyperreactive airway disease, and documentation of neurological deficits. Intraoperative concerns include safe manipulation of the airway during mask ventilation and the use of a flexible fibreoptic bronchoscope during endotracheal intubation to prevent neck flexion. It is also essential to avoid drugs, leading to histamine release. The use of multimodal monitoring including bispectral index and neuromuscular monitoring is crucial to prevent delayed recovery. Anticipation and management of exaggerated hypotension in response to anesthetic induction agents and prone position is the key to a successful outcome in patients with autonomic dysfunction.

Keywords: Hirayama disease, Monomelic amyotrophy, Autonomic dysfunction, Spastic quadriparesis

INTRODUCTION

Hirayama's disease, also known as monomelic amyotrophy prevalent in young males of Asian origin, is characterized by progressive, asymmetric atrophy of distal upper limbs muscles, in the absence of the involvement of the pyramidal tract.[1,2] The progression to autonomic dysfunction, pyramidal tract involvement, and quadriparesis are rarely reported. Given the dearth of literature regarding perioperative concerns, we report the anesthetic management of a case of Hirayama disease with autonomic dysfunction and progressive spastic quadriparesis undergoing trans-facetal C3–C7 screw fixation.

CASE REPORT

An adolescent male presented with gradual progressive weakness of the right forearm and hand followed by involvement of the left upper limb over a span of 4 years. He also developed weakness and stiffness in both lower limbs associated with exacerbations during winter (cold paresis). The patient also suffers from constipation and irregular coarse tremors on finger extension (minipolymyoclonus), for the past 6 months. He had an associated history of allergic rhinitis during winters.

The neurological assessment revealed wasting of the muscles of the forearm (right > left) with partial preservation of the brachioradialis (oblique atrophy sign), [Figure 1a] and atrophy of intrinsic muscles of both hands [Figure 1b]. The presence of spasticity and hyperactive deep tendon reflexes (DTRs) was noted in both lower limbs, and Babinski's sign was positive. His extremities were cold with a lack of hair on the dorsum of the hand indicating localized autonomic dysfunction.

Bedside tests of autonomic function showed the absence of orthostatic hypotension and <15 mmHg increase in diastolic blood pressure in response to isometric handgrip. Decreased amplitude of the compound muscle action potential of bilateral ulnar and median nerves was observed during the nerve conduction study. Needle electromyography of the triceps [Figure 1c] and abductor digiti minimi showed high amplitude, polyphasic action potentials, and reduced recruitment. Dynamic magnetic resonance imaging (MRI) revealed anterior displacement of the posterior dural wall during flexion and spinal cord compression at C3–C6 levels along with a crescent-shaped enhancement of the posterior epidural space (Crescent moon sign), [Figure 2]. The patient was then posted for C3–C7 trans-facetal screw fixation.

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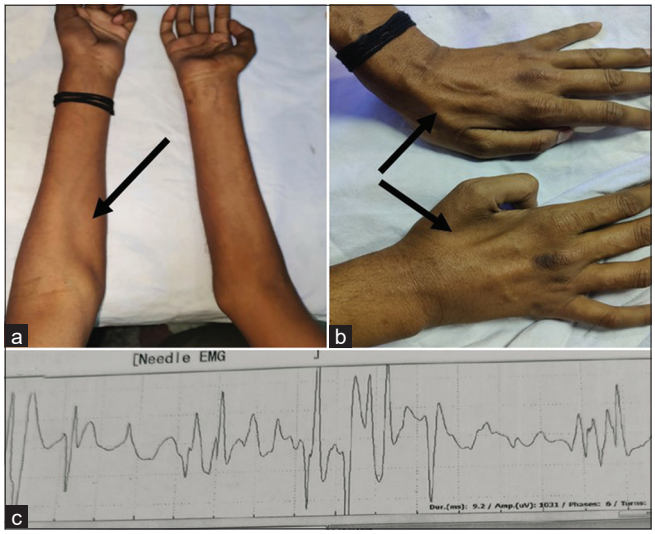

Figure 1: (a) Atrophy of the muscles of the forearm with preservation of the brachioradialis (Oblique atrophy sign) (black arrow). (b) Atrophy of the intrinsic muscles of the hand (thenar, hypothenar, and dorsal interosseous muscles) (black arrows). (c) Needle electromyography of the triceps revealing high-amplitude, polyphasic action potentials, fibrillation potentials, and reduced recruitment.

Inside the operation theatre room, standard monitoring along with bispectral index (BIS) and neuromuscular monitoring was instituted. The plan of airway management was flexible fiberoptic bronchoscope-guided (FOB) intubation under anesthesia. A preloading with 5 mL/kg crystalloid was followed by anesthetic induction using fentanyl (2 μg/kg), propofol titrated to loss of verbal response, and rocuronium (1 mg/kg) after confirmation of adequate bag and mask ventilation. FOB-guided endotracheal intubation was done. There was one episode of hypotension (90/60 mmHg) immediately post-induction which was managed with a single bolus of intravenous phenylephrine (100 μg). Invasive arterial monitoring was initiated and normovolemia was confirmed. Compression stockings were applied and the patient was turned prone with the head in the neutral position. There was a 2nd episode of hypotension (80/40 mmHg) immediately on turning prone which was managed with fluid bolus and phenylephrine infusion was started. The intraoperative period was uneventful and phenylephrine infusion (0.15–1.5 μg/kg/min) was tapered to stop by the end of the procedure. The patient was turned supine and extubated with a cervical collar *in situ*. He was discharged on post-operative day 5 with instructions to continue physiotherapy.

DISCUSSION

Hirayama disease was initially described as "juvenile muscular atrophy of unilateral upper extremity."[1] Our patient satisfied a majority of the criteria elucidated by Tashiro *et al*.; however, lower limb spasticity, increased DTR, and the presence of a positive Babinski's sign indicating an upper motor neuron (UMN) lesion were present.[2,3] Constipation (due to autonomic dysfunction) and UMN lesions are uncommon clinical manifestations of Hirayama disease.

Das and Pradhan evaluated the autonomic function in 44 patients with Hirayama and found decreased expiration/inspiration ratio of the heart rate (54.5%), 30/15 ratio of the R-R interval (60.6%), and acceleration-deceleration difference in response to standing (63.6%) revealing parasympathetic dysfunction, along with increased localized sweating (38.6%) denoting sympathetic overactivity.[4] Parasympathetic dysfunction is also associated with gastroparesis and reflux which increase the aspiration risk. Therefore, premedication should include an antacid and prokinetic and the patient should have adequately fasted.

Polyminimyoclonus, one of the hallmarks of the disease is defined as low-amplitude, intermittent, and arrhythmic movements accentuated when the hand is outstretched in extension.[2] This disease is diagnosed on the basis of the typical clinical manifestations and the crescent moon sign seen in dynamic flexion MRI.[2,3]

The pathophysiological basis of this disease is cervical flexion myelopathy secondary to the growth imbalance of the vertebral canal and dural canal resulting in compression of the spinal cord during flexion of the neck.[2] Hence, airway management as well as surgical positioning in the neutral position is deemed preferable. Meticulous care in the padding of pressure points and positioning should be taken in the presence of muscle atrophy and spasticity.

Hirayama *et al*. demonstrated circulatory insufficiency leading to necrosis of the alpha motor neuron cells of the spinal cord.[5] Hence, adequate preloading, use of compression stockings, and ensuring normovolemia using goal-directed fluid therapy along with maintenance of mean arterial pressure above 90 mmHg are essential to maintain adequate spinal cord perfusion. The use of direct-acting alpha-adrenergic agents for hypotension is preferred in the background of autonomic dysfunction. These patients are also prone to hypothermia and have exacerbated hypotension with blood loss during surgery. Ito *et al*. reported the presence of coexistent airway allergies with eosinophilia in patients with Hirayama.[6] Therefore, administration of drugs leading to histamine release (e.g., atracurium) should be avoided. Succinylcholine has the potential to cause hyperkalemia while long-acting muscle relaxants are associated with a prolonged neuromuscular blockade. Judicious use of anesthetic agents guided by BIS and neuromuscular monitoring is essential to prevent delayed recovery. Post-operative concerns include adequate analgesia, antiemesis, and physiotherapy facilitating early mobilization while reducing the risk of deep vein thrombosis and recurrent chest infections.

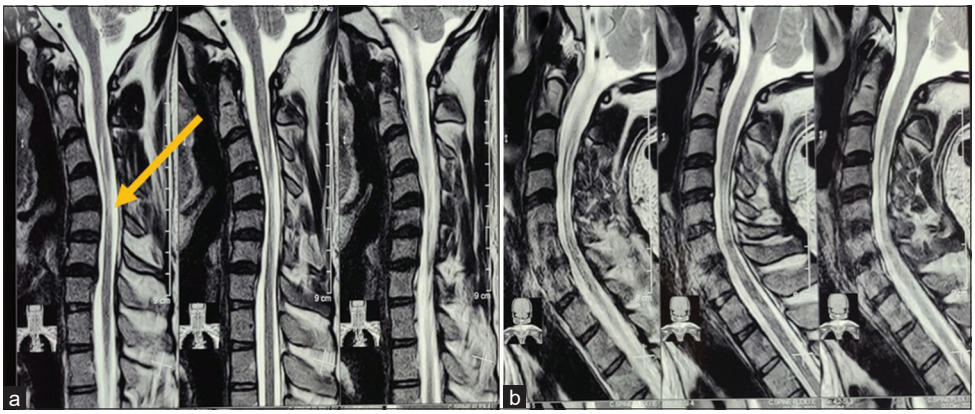

Figure 2: (a) Sagittal T2-weighted flexion magnetic resonance imaging (MRI) image demonstrating the anterior displacement of the posterior dural wall along with spinal cord compression and enlargement of the posterior epidural space (Crescent moon sign) (yellow arrow). (b) Sagittal T2-weighted extension MRI image demonstrating the presence of abundant cerebrospinal fluid anterior to the spinal cord and no spinal cord compression.

CONCLUSION

Hirayama disease can also present with uncommon features such as autonomic dysfunction and UMN lesion signs. The role of the anesthesiologist is crucial in the safe manipulation and securing of the airway, positioning, maintenance of stable hemodynamics, and successful extubation during trans-facetal cervical spine fixation surgery.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using the AI.

REFERENCES

1. Hirayama K, Toyokura Y, Tsubaki T. Juvenile muscular atrophy of unilateral upper extremity: A new clinical entity. Psychiatr Neurol Jpn 1959;61:2190-7.
2. Wang H, Tian Y, Wu J, Luo S, Zheng C, Sun C, *et al*. Update on the pathogenesis, clinical diagnosis, and treatment of Hirayama disease. Front Neurol 2022;12:811943.
3. Tashiro K, Kikuchi S, Itoyama Y, Tokumaru Y, Sobue G, Mukai E, *et al*. Nationwide survey of juvenile muscular atrophy of distal upper extremity (Hirayama disease) in Japan. Amyotroph Lateral Scler 2006;7:38-45.
4. Das A, Pradhan S. Cardiovascular and sudomotor dysfunction in Hirayama disease. Acta Neurol Belg 2021;121:545-53.
5. Hirayama K, Tomonaga M, Kitano K, Yamada T, Kojima S, Arai K. Focal cervical poliopathy causing juvenile muscular atrophy of distal upper extremity: A pathological study. J Neurol Neurosurg Psychiatry 1987;50:285-90.
6. Ito S, Kuwabara S, Fukutake T, Tokumaru Y, Hattori T. HyperIgEaemia in patients with juvenile muscular atrophy of the distal upper extremity (Hirayama disease). J Neurol Neurosurg Psychiatry 2005;76:132-4.

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