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Hereditary Neuropathy with Liability to Pressure Palsy and Vocal Cord Paralysis After Pulmonary Lobectomy: A Case Report

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Hereditary neuropathy with liability to pressure palsy (HNPP) is a rare peripheral neurological disorder that manifests with increased sensitivity to pressure. In people with this disorder, the peripheral nerves are unusually sensitive to pressure. Minor trauma or compression causing paralysis in the extremities is a hallmark of this disorder. Ensuring there is no pressure on the extremities is recommended as a preventive measure. We describe for the first time, postoperative vocal cord paralysis in a patient with HNPP due to left recurrent laryngeal nerve palsy. Anesthesiologists and surgeons should be aware of this possible complication in patients with HNPP. (A&A Practice. 2024;18:e01752.)

Hereditary neuropathy with liability to pressure palsy (HNPP) is a rare demyelinating peripheral neuropathy.[1] The symptoms of HNPP may include sensory and motor changes such as numbness, tingling, and muscle weakness in the limbs.[1] In most cases, it presents as acute mononeuropathy, which resolves within days to weeks, but can be recurrent and persistent.[1] The onset is usually at 20 to 30 years old, but cases in neonates and elderly patients have also been reported.[1] The primary management of HNPP is to avoid prolonged activity or posture to prevent peripheral nerve compression or stretch injury.[1] The nerves most affected by the disease are the peroneal, median and ulnar nerves, and the brachial plexus.[1] Previous studies have also demonstrated conduction abnormalities,[1] where pathological examination of nerve biopsy has shown sausage-like swelling of myelin sheaths, called "tomacula."[1] In most cases, chromosome deletion is observed in the 17p11.2-p12 region, including PMP22.[1]

In previous cases, anesthesia-related neuropathy in HNPP has mostly involved the peripheral nerves of the extremities.[2–12] It has been recommended that any pressure involving the extremities be minimized during anesthesia to prevent these complications. We report a case that successfully managed to prevent peripheral nerve neuropathy, but the patient manifested with vocal cord paralysis due to recurrent laryngeal nerve (RLN) palsy. Written informed consent for publication was obtained from the patient.

CASE DESCRIPTION

A 69-year-old, height 162 cm and weight 66 kg, woman with a history of HNPP was admitted for a video-assisted left upper lobectomy for lung carcinoma. Her HNPP was diagnosed several years before. At that time, the patient exhibited an inability to stand up after crossing her legs for a short period. She also had pain and numbness in her left shoulder. A nerve conduction velocity test revealed a decrease in speed of electrical impulses in the left radial nerve, and a nerve biopsy showed demyelinating changes. Furthermore, genetic analysis showed a deletion of chromosome 17q11.2 (PMP22). During her preoperative examination, her symptoms were sensory abnormality only: paresthesia in her left arm. Preoperatively, neurology was consulted, and they recommended careful positioning during surgery. The day before surgery, the patient, anesthesia, surgery, and nursing team met and conducted a simulation of positioning and checked all pressure points.

In addition to the standard monitors, a radial arterial catheter was placed in the operating room under ultrasound guidance for blood-pressure measurements. General anesthesia was induced with propofol, fentanyl, and remifentanil. An electromyographic neuromuscular blockade monitor (AF-200, Nihon Koden Co.) was set and calibrated, and then rocuronium was administered. Train-of-four (TOF) monitoring was started with 5-minute intervals between stimulations. After confirming the TOF count at zero, a 35Fr. left-sided double-lumen endotracheal tube (ETT) was inserted without incident using a video laryngoscopy (McGRATH MAC3, Medtronic Japan Co, Ltd.). The glottic view was grade II based on the Cormack-Lehane classification. The position of the ETT was verified using a fiberscope, and it was taped at the right corner of the mouth. The cuff pressure was within normal range at 26 cm H_2O. After intubation, the patient was placed in a right lateral decubitus position for surgery. Careful attention was given to all pressure points, and pressure relief

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pads were placed under the extremities. Pressure point checks were performed every 30 minutes during surgery. The course of the surgery was uneventful. The patient was extubated in the operating room and transferred to the recovery room. The duration of surgery was 3 hours and 22 minutes, and the total time in the operating room was 4 hours and 26 minutes.

Postoperatively, the patient did not exhibit any new or worsening symptoms of her neuropathy. However, she felt difficulty in speaking but did not complain of it to the medical staff. As there was no worsening of the symptoms, the patient was discharged on postoperative day 6. After 1 month, she could speak but was having difficulty singing, which severely affected her profession as a troubadour of traditional Chinese poems. The patient started voice rehabilitation in an otorhinolaryngology clinic, but there was no significant improvement. Seven months after surgery, a flexible fiberoptic laryngoscopy examination revealed vocal cord paralysis due to left RLN paralysis. Subsequently, she was treated with a Fibroblast Growth Factor injection to the vocal folds to restore function.

DISCUSSION

Thus far, there have been 11 case reports of patients with HNPP who underwent surgery or procedure (Table).[2–12] The types of anesthesia were general anesthesia (n = 5),[5,8–10,12] epidural anesthesia (n = 1) for cesarean delivery,[4] epidural analgesia for labor (n = 5),[2,3,6,11] and spinal anesthesia for Cesarean delivery (n = 3).[3,7,12] Four patients had no neurological complications,[2,3,11,12] whereas 9 experienced some complications.[4–12] The condition of 6 patients improved within 2 days to 5 months.[4,5,8,9,11,12] In 2 patients, nerve injury persisted.[7,10] One patient recovered completely; however, no details were described.[6]

Among the neuropathies reported for patients with HNPP who had surgery, cranial nerve injury is rare as an outcome. Although the details were not provided, a case of postoperative tongue dysesthesia after general anesthesia has been reported.[12] To the best of our knowledge, this is the first case report involving postoperative vocal cord paralysis. There is a report of nonsurgical vocal cord paralysis in a 19-year-old female with HNPP. She developed hoarseness and dysphasia after sleeping in the prone position.[13] The authors attributed these symptoms to right vocal cord paralysis by pressure or compression. Her symptoms resolved completely in 6 weeks.

In our case, vocal cord paralysis was due to left RLN paralysis. Multiple factors have been suggested for RLN paralysis after general anesthesia in the literature,[14,15] including hyperextension of the neck, inadequate ETT size or overinflated cuff, surgical injury, and compression by a gastric tube. Since we positioned the patient very carefully and kept her neck in a neutral position, hyperextension of the neck was an unlikely cause. We did not insert a gastric tube. Therefore, we hypothesized 2 possible causes for this: intubation with a double lumen ETT and/or lymph node dissection during surgery. Although the size of ETT was adequate and the cuff pressure was within a normal range, it is still possible that the ETT compressed the left glottis and larynx. As Morgen and Figueroa[10] discussed in their case report, 1 should consider securing an ETT at the center of the mouth, and properly supporting the ETT and corrugated respiratory circuit so they don't exert pressure against the patient's lips, glottis and larynx. Our surgeons denied the possibility of intraoperative RLN injury; thus, the cause of vocal cord paralysis cannot be determined.

ANESTHETIC CONSIDERATIONS IN PATIENTS WITH HNPP

Positioning

Since trivial insult can cause nerve injury, the primary preventive measure is minimizing the impact of pressure, compression and stretch. Special considerations should be given to any possible sites of neuropathy. Morgan and Figueroa[10] recommended simulating the surgical position in an awake state to ensure proper positioning. In addition, they recommended that the patient's arm be positioned to the side (<90°) and that the arm be rotated (supinating/pronating) every 15 minutes to prevent brachial plexus injury.[10] Bolger and Stewart[11] reported a case of a patient with HNPP who had labor and delivery with epidural analgesia. The patient

Table. Postoperative Neuropathy in Patients With Hereditary Neuropathy With Liability to Pressure Palsy

Literature	Surgery	Anesthesia	Position	Nerve	Outcome
Lepski and Alderson[2]	Labor and delivery	Epidural	n.d.	Brachial plexus	Healing
Berdai et al[3]	Labor and delivery	Epidural	n.d.	None	
	Elective C/S	Spinal	n.d.	None	
Peters and Hinds[4]	Emergent C/S	Epidural	n.d.	Peroneal n.	5 mo
Wijayasiri et al[5]	Mastectomy	General	supine, arm abduct 90°	Radial n.	4 wk
Llácer Pérez et al[6]	Labor and delivery	Epidural	n.d.	Peroneal n.	Healing
Chilvers and Salman[7]	Emergent C/S	Epidural	n.d.	Low back sensory disturbance	No healing
Kramer et al[8]	Total knee arthroplasty	General	n.d.	Peroneal n.	2 mo
Logroscino et al[9]	Hip prosthesis revision	General	n.d.	Peroneal n.	3 mo
Morgan and Figueroa[10]	Firs rib resection (thoracic outlet syn.)	General	decubitus	Peroneal n.	No healing
Bolger and Stewart[11]	Labor and delivery	Epidural	n.d.	Peroneal n.	3 mo
	Labor and delivery	Epidural	n.d.	None	
Samuel et al[12]	n.d.	General	n.d.	Tongue paresthesia	n.d.
				Loss of arm sensation	2 d
	Elective C/S	Spinal	n.d.	None	

Abbreviations: C/S, cesarean delivery; epidural, epidural analgesia or anesthesia; general, general anesthesia; n., nerve; n.d., not described; nerve, injured nerve; outcome, the outcome of postoperative neuropathy indicates good outcome (healing) or no healing, and if healing took time, time to heal is listed; position, intraoperative position; spinal, spinal anesthesia; syn., syndrome.

experienced a common peroneal nerve paralysis after her delivery, which required 3 months of recovery. After being diagnosed with HNPP, the patient became pregnant again. Several preventive measures were taken during the second labor and delivery with epidural, and the patient did not experience neuropathy. Thus, postsurgical neuropathy may be preventable by careful positioning and taking appropriate preventive measures.

Arterial Line

Samuel et al[12] recommend using an arterial line for blood-pressure monitoring instead of a blood-pressure cuff to avoid medial or ulnar nerve injury. We placed a radial arterial line under ultrasound guidance to reduce the chance of unsuccessful insertion or complications such as hematoma formation.

Intermittent Pneumatic Compression Device

There is a concern that an intermittent pneumatic compression device for preventing deep venous thrombosis may cause pressure injury.[12] In our case, the compression device was used after the risks and benefits were discussed with the patient.

Neuromuscular Monitoring

The typical electrophysiological feature observed in HNPP is the reduction of motor nerve conduction velocities with prolonged distal motor latencies.[1] Therefore, monitoring of muscle relaxation and careful TOF calibration (eg, every 5 minutes, set the first stimulus current at 30 mA, and optimal stimulus current of 60 mA) may be a valuable tool to monitor and detect undiagnosed HNPP.

In summary, we experienced a case in which preventive measures were implemented in a patient with HNPP who underwent a lung resection. The patient exhibited no new or worsening of peripheral neuropathy but the postoperative course was complicated by unexpected postoperative vocal cord paralysis. Currently, neuropathy in HNPP during anesthesia is mainly focused on the extremities. However, the RLN is also vulnerable to pressure and minor trauma. Therefore, anesthesiologists and surgeons should be aware of this potential neuropathy of the RLN in patients with HNPP.

DISCLOSURES

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