

Carotid Endarterectomy for a Patient with a Right-sided Aortic Arch and Aberrant Left Subclavian Artery Predicting a Left Non-recurrent Inferior Laryngeal Nerve: A Case Report and Literature Review

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Abstract

Cardiovascular malformations during embryogenesis can lead the inferior laryngeal nerve to branching directly from the cervical vagus nerve and entering the larynx. This rare anatomical variation is known as a non-recurrent inferior laryngeal nerve (NRILN), and increases the risk of accidental injury resulting in postoperative vocal cord paralysis during neck surgery. We report a case of an 83-year-old man who presented with left symptomatic internal carotid artery stenosis with a right-sided aortic arch and aberrant left subclavian artery (ALSCA). We performed carotid endarterectomy (CEA) using intraoperative neuromonitoring to avoid NRILN injury. To the best of our knowledge, this is the first report of searching for a left NRILN by electrophysiology during CEA. Neurovascular surgeons need to understand the variations of the NRILN associated with congenital cardiovascular anomalies and effective use of intraoperative nerve monitoring (IONM). We discuss the embryological origin, IONM, and surgical pitfalls of this case.

Keywords: aberrant subclavian artery, carotid endarterectomy, intraoperative nerve monitoring, non-recurrent inferior laryngeal nerve, right-sided aortic arch

Introduction

Accurate knowledge of anatomical variations associated with the carotid arterial system is essential for safe and reliable procedures during carotid endarterectomy (CEA). The inferior laryngeal nerve is the terminal branch of the recurrent laryngeal nerve (RLN) arising from the vagus nerve in the superior part of the thorax.¹⁾ The RLNs that control the function of vocal cords are asymmetrical branches of the vagus nerve, looping under the right subclavian artery on the right side and the aortic arch on the left, and ascending into the larynx. Because of an embryological error, the RLN can originate from the cervical vagal trunk and travel directly into the larynx. This is called the non-recurrent inferior laryngeal nerve (NRILN). The right NRILN with an aberrant right subclavian artery (ARSCA) has an

incidence of 0.7%.^{2–6)} The left NRILN is extremely rare with an incidence of approximately 0.04% because of its association with a right-sided aortic arch and aberrant left subclavian artery (ALSCA).^{3,4,7,8)} This abnormality is well known in thyroid and parathyroid surgeries and has a higher surgical risk of injury compared with normal anatomy.^{2,5)} Neurovascular surgeons should also recognize the features of the NRILN and not impair this anatomical variation during exposure of the carotid artery. To date, however, limited reports on CEA with an NRILN have been published.^{7–10)} The present report describes a patient with left internal carotid artery stenosis accompanied by a right-sided aortic arch and ALSCA. CEA was successfully performed with intraoperative nerve monitoring (IONM) for a left NRILN. The surgical significance, IONM, and embryological origin of this anomaly are discussed.

Case Report

An 83-year-old man presented with mild monoparesis of right lower extremity and suffered from left

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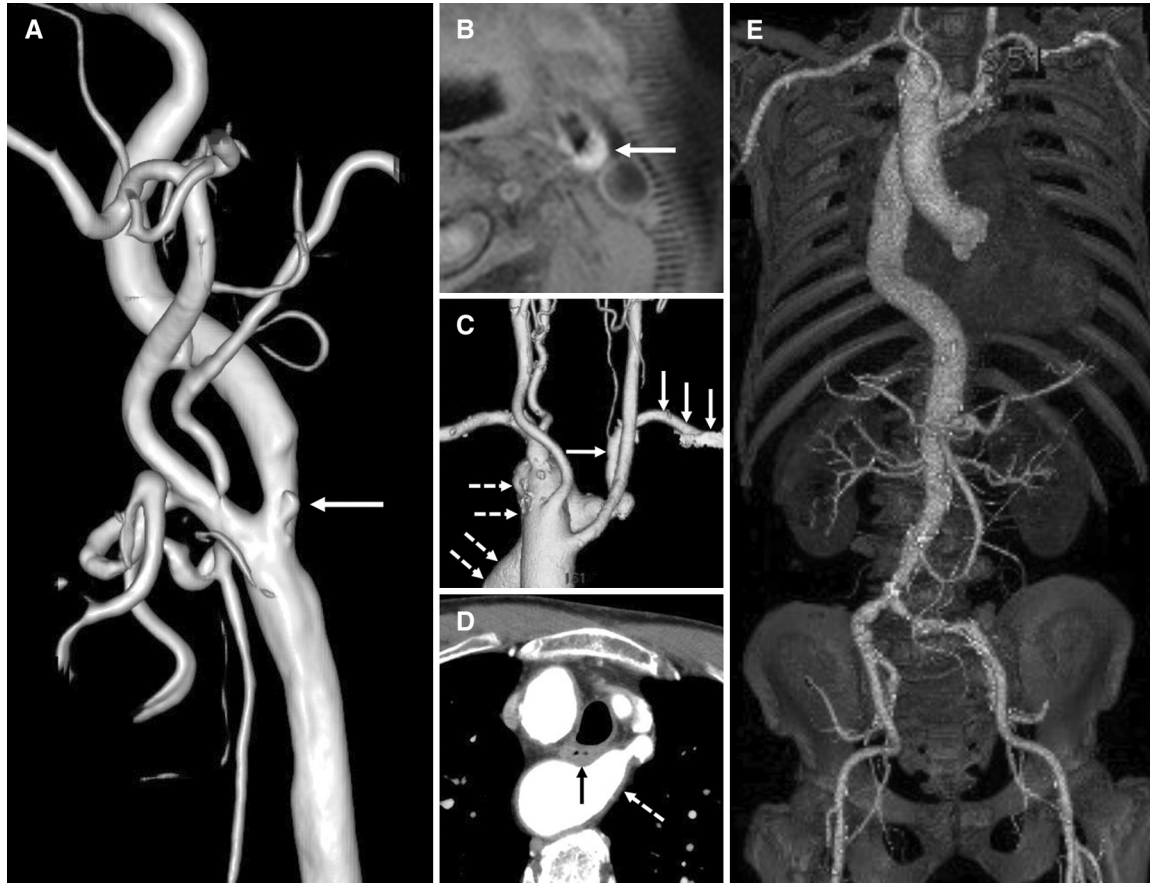


Fig. 1 Preoperative vascular evaluation. (A) Lateral view of the left carotid 3D digital subtraction angiography revealed a 60% stenosis with the ulcer (white arrow) at the internal carotid artery. (B) Axial view of T1-weighted magnetic resonance carotid plaque imaging showed hyperintensity (white arrow), suggesting an unstable plaque. The signal intensity ratio was 1.54. (C) 3D CTA demonstrated an ALSA (white arrows) originated from a right-sided aortic arch (dotted white arrows) as a fourth branch of the aortic arch. (D) CTA depicted Kommerell's diverticulum (dotted white arrow), the origin of the ALSA. The ALSA coursed posterior to the esophagus (black arrow). (E) 3D reconstruction of CTA indicated the heart located in a normal position. 3D: three-dimensional, ALSA: aberrant left subclavian artery, CTA: computed tomography angiography.

cerebral infarction. He was diagnosed with moderate atherosclerotic stenosis at the origin of the left internal carotid artery (Fig. 1A). The mechanism underlying the infarction appeared to be artery-to-artery embolism from this stenosis. The carotid ultrasound showed echolucent plaques, a marker of plaque vulnerability. Unstable plaques with intraplaque hemorrhage were also indicated due to high-intensity on T1-weighted imaging (Fig. 1B). The computed tomographic angiography showed a right-sided aortic arch, retroesophageal ALSA, and Kommerell's diverticulum without situs inversus (Figs. 1C–1E).

To prevent recurrent stroke, surgical treatment for carotid artery stenosis was recommended. Treatment was selected based on plaque vulnerability and the treatment risk evaluation. The presence of a right-sided aortic arch with an ALSA suggested the

possibility of a left NRILN. CEA has an additional risk of NRILN injury. In contrast, unfavorable complex vascular anatomy posed a challenge of access into the left carotid artery and a risk of Kommerell's diverticulum injury. CEA was recommended with the use of IONM to detect and identify the NRILN in addition to routine intraoperative monitoring. Left CEA was undertaken with the patient under general anesthesia. A neural integrity monitor (NIM) electromyogram (EMG) endotracheal tube (NIM EMG Endotracheal Tube; Medtronic Xomed, Jacksonville, FL, USA) was appropriately positioned between the vocal cords using a bronchoscope. Before the identification of the vagus nerve, a monopolar probe (NIM accessory; Medtronic Xomed) with current of 2 mA was used for initial searching out the NRILN. While dissecting tissue near the carotid artery, direct

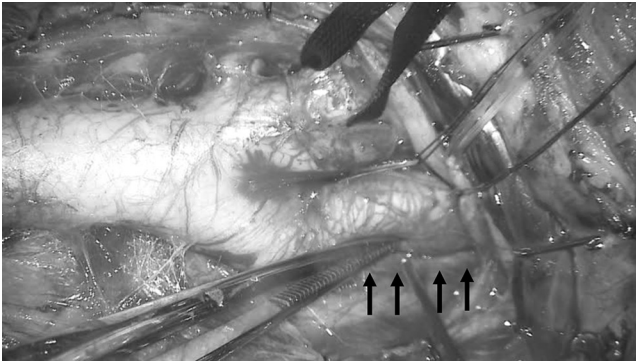


Fig. 2 Intraoperative photograph during left CEA. The vagus nerve (black arrows) was exposed. The proximal segment of the vagus nerve was stimulated by a monopolar probe with an electrical current. Endotracheal tube-based system recorded positive EMG signals. CEA: carotid endarterectomy, EMG: electromyogram.

and frequent checking by the NIM system (NIM-Response 3.0 system; Medtronic Xomed) on the dissection site showed no EMG signal. After exposure of the vagus nerve, lying between and just posterior to the common carotid artery and internal jugular vein, the proximal vagal stimulation at 0.8 mA got positive EMG signals (Fig. 2). After the visual and electrophysiological identification of the vagus nerve, its nerve was stimulated proximally to distally, and good EMG response was obtained in all positions. This pattern of both proximal and distal vagal stimulation with the EMG response confirmed the absence of a NRILN in the surgical field. CEA was completed with no considerable changes in routine intraoperative monitoring.

Postoperative imaging studies showed no stroke. No cranial nerve palsy was found, and normal vocal cord function on direct laryngoscopy was confirmed. The patient's postoperative course was unremarkable.

Discussion

RLN palsy is one of the well-known complications in neck surgery.^{2,5,11,12} This results in hoarseness and dysphasia, and it induces glottal obstruction and serious airway compromise when bilateral nerves are injured. NRILN is a rare anatomical variation and represents a major surgical risk of iatrogenic injury during thyroid and parathyroid surgery because of unawareness of its presence.^{2,5} After the first report on the right NRILN with an ARSCA by Stedman in 1823,¹³ several reports have established the clinical significance of this abnormal nerve in the literature.

To comprehend the anatomy of the NRILN, involution and embryological errors of the primitive six aortic arch pairs, termed branchial arches between the ventral and dorsal aorta, need to be understood. Transformation and regression of branchial arches in the developing embryo result in formation of the RLN, with the right nerve looping under the right subclavian artery and the left nerve looping under the aortic arch and ductus arteriosus (DA) (Fig. 3A).^{14–17} The right NRILN is associated with anomalous regression of the right fourth arch (right subclavian artery), which causes an ARSCA originating from the aorta distal to the left subclavian artery. As a result, the right RLN is no longer hauled by the right fourth branchial arch, and then it migrates upward and becomes non-recurrent (Fig. 3B). In the comprehensive meta-analysis, 86.7% of patients with right NRILN showed an ARSCA pattern.⁶ The left NRILN requires more complex regression of the left fourth arch (adult aortic arch) and sixth arch (DA). According to the Edwards' classification of the double aortic arch system, the development of a left NRILN requires Edward's Type II regression, which involves the right-sided aortic arch with ALSA.^{17,18} A left NRILN also necessitates a right DA. In the case of a right-sided aortic arch and ALSA with situs inversus, the DA exists on the right side and the left inferior laryngeal nerve takes a non-recurrent course (Fig. 3C).^{3,14–17} With regard to a right-sided aortic arch and ALSA without situs inversus, Kommerell's diverticulum, an aortic dilatation from which the aberrant subclavian artery originates, may be the connection point with the DA.^{14,15,17} However, radiological detection of DA is difficult because it becomes a narrow ligament after birth, and the incidence of a right DA has not been clarified. Therefore, a right-sided aortic arch with an ALSA should be regarded as an indication of a potential left NRILN.

Natural history of Kommerell's diverticulum is unknown due to its rareness.¹⁷ Some investigators have been reported rupture rate of 6–19% among affected patients and high mortality rate with the rupture.^{19,20} High dissection rate also has been shown.^{20–22} Although the reported orifice size of the dissected and/or ruptured diverticulum ranged from 2 to 10 cm,^{19–22} the safe state without the rupture cannot be predicted because of its rarity. When guiding catheter and guidewire are navigated into the left common carotid artery in neurovascular treatment, careless advancement of the devices might result in the injury of Kommerell's diverticulum and death.

The RLN normally ascends posterior to the carotid artery, and its course is located medially in the dissected field of CEA.^{11,12} The NRILN travels in another pass

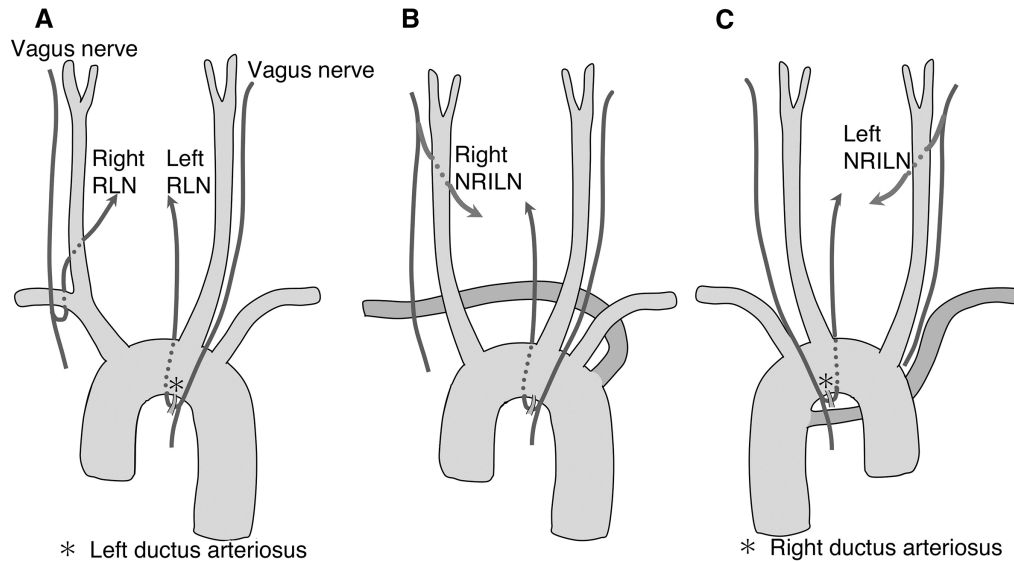


Fig. 3 Anatomical courses of the RLN and NRILN. (A) Normal pathway of the RLN. (B) The anomalous origin of the right subclavian artery from a point distal to the left subclavian artery results in the right NRILN. (C) An ALSA and right-sided aortic arch with the right ductus arteriosus cause the left NRILN. ALSA: aberrant left subclavian artery, NRILN: non-recurrent inferior laryngeal nerve, RLN: recurrent laryngeal nerve.

around the carotid artery. Toniato et al. classified the NRILN pathway (Fig. 4).⁵⁾ NRILN type 1, which is observed most frequently with an incidence of 58.3%,⁶⁾ originates above the laryngotracheal junction and runs together with a superior thyroid artery. NRILN type 2 follows a transverse path parallel to an inferior thyroid artery. When NRILN type 1 crosses anterior to the carotid artery, careless exposure in CEA is likely to damage this nerve.^{7,8)} The origin of the NRILN is not detectable using preoperative investigations. Additionally, six cases of the NRILN without vascular anomalies have been reported and four of these cases had an NRILN accompanied by an ipsilateral RLN.^{4,7,23,24)} Even though preoperative imaging indicates the absence of large vascular abnormalities, the NRILN should always be considered during surgical neck exploration. There is another potential problem with the NRILN. The communicating branches between the cervical sympathetic system and the RLN have a similar diameter as the NRILN with an incidence of 1.5%.^{1,25)} These nerves can be mistaken for the NRILN.

Electrophysiological IONM helps identify the RLN as the gold standard during thyroid and parathyroid surgery.²⁶⁾ An endotracheal tube sensing vocal cord motion, such as an EMG endotracheal tube, is used at intubation. The NIM system records EMG data from the vocal cords with audio and visual waveform information regarding evoked waveforms. When an NRILN exists near the dissection point, the NIM system indicates the EMG response and the surgeon should dissect the tissue more carefully and identify

the nerve visually. After opening the carotid sheath, the vagal trunk between the internal jugular vein and carotid artery is found.^{1,11)} The vagus nerve is stimulated proximally to distally, and the NRILN separates from the vagal trunk at the point where the NIM system decreases EMG signals (Fig. 4).^{27,28)} Standardization of IONM and strict criteria of troubleshooting algorithm enable to increase positive predictive value (75% or higher) and negative predictive value (92–100%).^{26,29)} The dominant source of false positives (i.e. loss of signal with intact vocal cord function) is malposition of the endotracheal tube. Reconfirmation of impedance values may allow to detect rotational tube displacement. Other causes of false-positive IONM results include the problems with the technical device, insufficient stimulating current, blood or fascia covering the stimulated nerve segment, delayed neuromuscular blockade reversal, and pooled saliva at the glottis. Postoperative vocal cord paralysis with false-negative scenarios (i.e., EMG response with the nerve injury) is often temporary without severe symptoms.²⁶⁾ To make visualization of the nerves easy and reduce the risk of IONM failure, the neurovascular surgeon must perform safe and secure hemostasis, especially during the CEA under the condition of antithrombotic therapy.

In the present case, an NRILN was not encountered intraoperatively. We concluded that the NRILN corresponded to Toniato's type 2 and followed a caudal pathway out of the dissecting field, or was absent (i.e., normal course of the left RLN). This is

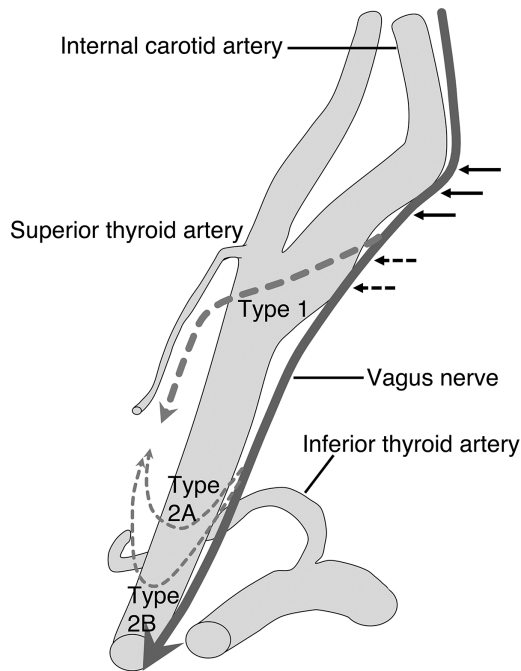


Fig. 4 IONM of the NRILN. Toniato's type 1 NRILN traverses near the carotid bifurcation. Toniato's type 2 NRILN crosses parallel to the inferior thyroid artery (over the inferior thyroid artery, type 2A; and under or between the inferior thyroid artery, type 2B). As an example of type 1 NRILN, a case of positive EMG response from the upper vagal stimulation (black arrows) and negative EMG response from the lower vagal stimulation (dotted black arrows) indicate the separation of the NRILN. EMG: electromyogram, IONM: intraoperative nerve monitoring, NRILN: non-recurrent inferior laryngeal nerve.

the first report of searching a left NRILN by electrophysiological monitoring during CEA with a right-sided aortic arch and ALSA.

Preoperative recognition of an aberrant subclavian artery leads to prediction of an NRILN. When an NRILN is predicted, neurovascular surgeons should use IONM, and pay close attention to the NRILN during carotid artery exploration. Additionally, sporadic NRILN without vascular abnormalities can exist. Careful dissection and detailed knowledge of normal and aberrant anatomy allow for avoidance of nerve injury during CEA.

Conflict of Interest Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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