

## Neurilemmomas of the Cervical Vagus Nerve

Cheong Soo Park, M.D., Kwang Wook Suh, M.D. and Choon Kyu Kim, M.D.

*Department of Surgery, Yonsei University College of Medicine, Seoul, Korea*

### 경부 미주 신경에 발생한 신경초종

연세대학교 의과대학 외과학교실

박정수 · 서광욱 · 김춘규

=국문초록=

인체에서 발견되는 전체 신경초종중 25~40%가 두경부에서 발견되나 경부미주신경의 신경초종은 매우 희귀하여 1988년까지 영문문헌상 88예가 보고되어 있다. 임상적으로 경부종괴가 주증상이며 때로는 애성, 종괴촉지시 기침 혹은 방사통, parapharyngeal space의 종괴성장으로 연하곤란등이 초래되기도 한다. 치료는 종괴의 완전적출과 미주신경의 기능을 보존시키는 것이 주요 목표로 되어 있다.

저자들은 최근 5년간 4예의 경부미주신경의 신경초종을 경험하였는데, 3예는 측경부의 상부에, 1예는 측경부의 하부에 발생한 것이었다. 연령은 25세에서 50세까지 성인연령이었고, 성별은 남자 1예, 여자 3예이었다. 내원시 주소는 전예가 무통성 경부종괴이었으며, 종괴의 크기는 직경 3cm에서 10cm까지 다양하였다. 전예에서 종괴로 인한 신경학적 증상은 없었으나, 1예에서 parapharyngeal space의 거대종괴 때문에 다소간의 연하곤란이 있었다. 2예에서는 종괴촉지시 기침이 유발됨을 호소하였다. 수술은 경부횡절개로 흉쇄유돌근을 제치고 총경동맥과 내경정맥을 종괴의 상하부위가 완전히 노출되도록 박리한 후 종괴의 피막을 미주신경의 주행방향에 따라 절개하여 적출(enucleation)하였다. 적출술시 종괴가 유착되어 있는 피막 부위는 동시에 절제하되 미주신경의 신경경로(neural pathway)는 유지되도록 하였다.

수술적 후 2예에서 일시적 애성을 호소하였다. 추적은 최단 20개월에서 최장 80개월까지 하였는데 일시적 애성은 호전되었고, 전예가 재발없이 건강하게 지내고 있었다.

따라서 본 종양수술은 종양적출술시 이환된 미주신경의 절단을 결정하기 전에 신경경로를 유지시킬 수 있는 적출술(enucleation)을 먼저 고려하는 것이 바람직하다고 사료되었다.

**KEY WORDS :** Neurilemmoma · Vagus nerve.

Neurilemmoma is a benign tumor of Schwann cell origin and may occur on any nerve covered by Schwann cells<sup>1)2)</sup>. Although approximately 25

to 40% of all neurilemmomas occur in the head and neck region, the lesions originating from the cervical vagus nerve are extremely rare<sup>3)</sup>. To our

knowledge, until 1988, only 88 neurilemmomas of the vagus nerve had been reported in the literature<sup>4</sup>).

Clinically, the most common presenting symptom is a painless lateral neck mass, making it difficult to differentiate from other solid masses of the neck. Since these tumors are benign, the preferred treatment is a complete surgical extirpation with conservation of the neural pathway if at all possible.

The objectives of this paper are to present 4 case reports of cervical vagus neurilemmomas over a 5-year period and to review the clinical, pathologic, and surgical aspects of the lesions.

### Case Reports

#### Case 1 :

A 50-year-old woman was initially seen with an 18-month history of a slowly growing mass

in the left retromandibular space. Physical examination revealed a round, smooth-surfaced, non-tender mass measuring 3×3cm located in the lower part of the left retromandibular space. On palpating the mass, a paroxysmal cough was elicited, but no signs of nerve paralysis were found.

She underwent exploration of the left side of the neck through a transverse skin incision over the mass. A non-pulsatile, well-encapsulated mass arising in the vagus nerve was found between the internal jugular vein and the carotid bifurcation. An intracapsular enucleation of the tumor with a careful dissection of the splayed nerve trunk off the tumor by use of an operating microscope was accomplished, and the main trunk of the vagus and its branches were preserved. Frozen-section examination of the specimen was suggestive of a neurilemmoma.

Postoperatively, she had a left vocal cord palsy that gradually disappeared in 12 months. Histolo-

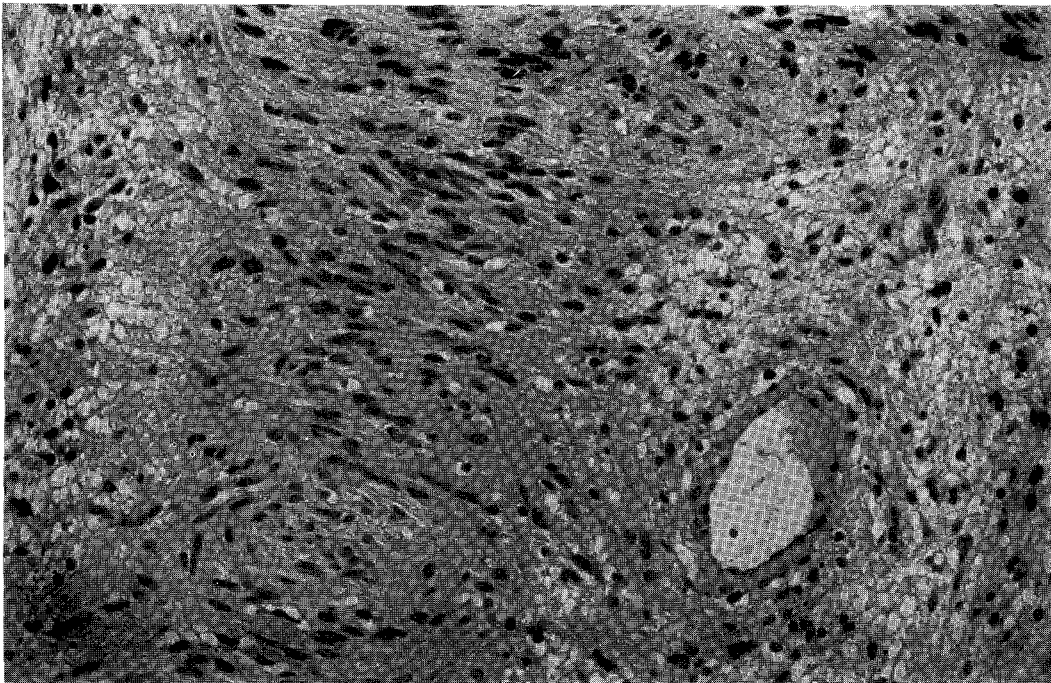


Fig. 1. Case 1 : Histologic section showing a mixture of Antoni type A and B tissues. Note the compact Schwann cells with nuclear palisading in the center of the field(Antoni A) and the loosely arranged reticulum network in the periphery of the field(Antoni B). Hematoxylin and eosin, ×150.

gic examination confirmed a typical neurilemmoma of Antoni type A and B mixed architecture (Fig. 1). The patient has remained free of the disease for 80 months following surgery.

#### Case 2 :

A 25-year-old woman was initially seen with a 12-month history of a growing mass in the high upper left lateral neck. The mass measured 8×4cm and was mobile horizontally but not vertically. The clinical diagnosis was a possible salivary gland tumor originating from the submaxillary gland.

At operation, a smooth-surfaced and shiny gray fusiform mass was found arising in the vagus nerve just below the posterior belly of the digastric muscle. Complete enucleation of the tumor was accomplished with preservation of the main trunk of the vagus nerve(Fig. 2).

Postoperatively, the patient demonstrated no vocal cord palsy and remained well without evidence of recurrence for 64 months after surgery. Histologic examination of the specimen confirmed a neurilemmoma with a mixed architecture of both Antoni type A and B tissues.

#### Case 3 :

A 29-year-old man was initially seen with a

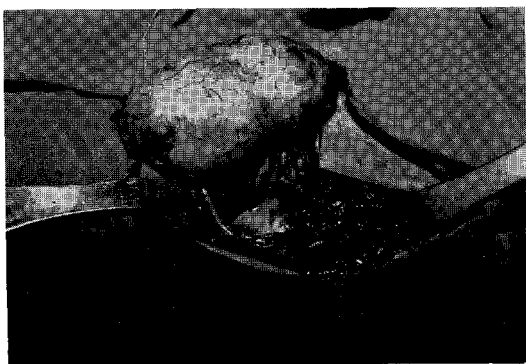


Fig. 2. Case 2 : Intraoperative view of the neurilemmoma enucleated from the cervical vagus nerve.



Fig. 3. Case 3 : Ultrasonography showing a mixed echogenic mass(M) between the carotid artery (A) and the internal jugular vein(V).

mass in the left lateral neck for 24 months. On palpating the mass, a paroxysmal cough was produced but demonstrated no neurologic deficits. Ultrasonography of the neck revealed a mixed echogenic mass located just beneath the left sternocleidomastoid muscle(SCM) and between the common carotid artery and the internal jugular vein(Fig. 3).

At operation, a 5×3cm well-encapsulated globular mass was found arising in the vagus nerve around the intersection of the superior belly of the omohyoid muscle and the SCM. By careful intracapsular dissection, the tumor was completely enucleated while preserving the main trunk of the vagus.

Postoperatively, he demonstrated a left vocal cord palsy that was gradually improved in 6 months. Histological appearance of the specimen was compatible with a neurilemmoma of Antoni type A and B mixed tissues. The patient has remained free of the disease for 25 months following surgery.

#### Case 4 :

A 49-year-old woman was initially seen with a 15-month history of a growing mass in the left neck and a 1-month history of mild swallowing difficulty. A neck computed tomographic(CT) scan with contrast showed a large oval mass occu-

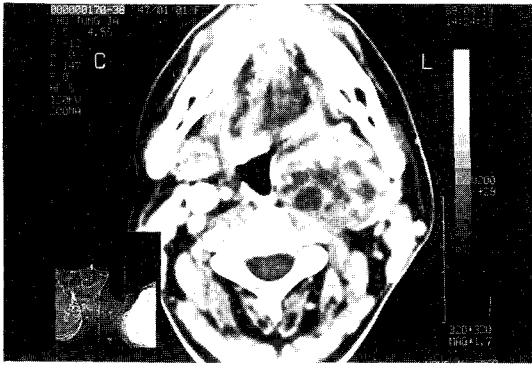


Fig. 4. Case 4 : Axial CT scan showing a low density mass occupying the left parapharyngeal space. Note a pronounced bulging into the oropharyngeal area.

pying the left parapharyngeal space with a pronounced bulge in the oropharyngeal wall (Fig. 4).

At operation, an encapsulated ovoid mass measuring  $10 \times 5 \times 5$  cm was found arising in the vagus nerve in the parapharyngeal space, and its consistency was that of a soft lemon. After longitudinal splitting of the nerve sheath splayed over the tumor surface, the tumor was enucleated by a careful intracapsular dissection. A small portion of the adherent fascicles, however, was included with the main specimen, because the adequate dissecting plane was not distinct in spite of the use of an operating microscope.

There was no paralysis of vocal cord postoperatively, and the patient remained well with no evidence of recurrence for 20 months after surgery. Histologic examination of the specimen confirmed a neurilemmoma of Antoni type A tissue predominant architecture, a so-called cellular neurilemmoma.

## Discussion

The most common location for vagal neurilemmomas is near the nodosa ganglion, but they may be found at any level along the course of the cervical vagus nerve<sup>4</sup>). These tumors usually appear as a lateral neck mass of insidious origin, and

may be frequently confused with paraganglioma, neurofibroma, malignant lymphoma, lipoma, branchial cleft cyst, metastatic cervical lymphadenopathy, submaxillary gland tumor, tumor of the retromandibular portion of the parotid gland, and tuberculous cervical lymphadenopathy<sup>5)6</sup>).

These tumors occur at any age, but the majority of patients have been in the third to fifth decades of life. There is no preponderance in sex incidence<sup>7</sup>). The mass is usually fusiform, ovoid, or spherical in contour, firm to rubbery hard in consistency, and can usually be moved horizontally but not vertically. On occasion, a coughing spell may be produced with pressure on the mass<sup>6)7)8</sup>). Large tumors may cause pressure symptoms within the confined anatomic regions and other symptoms such as dysphonia, dysphagia, and dyspnea. If the tumor is not removed, it can become enormously large and may undergo hemorrhage and cystic degeneration<sup>6)7)8</sup>).

Preoperative evaluation using ultrasound or CT scans of the neck may be helpful in determining the extent of the tumors and assist in the differential diagnosis of lateral neck masses. Recently, magnetic resonance imaging has been shown to be superior to CT scans for evaluating nerve origin tumors<sup>4)8-10</sup>). In our opinion, however, surgical excision and frozen-section examination appear to offer a better chance for correct diagnosis and effective treatment.

Histologically, the components of tumor cells have been classified into two discrete ones, Antoni type A and B tissues<sup>11</sup>). Antoni type A tissues are characterized by compact Schwann cells with nuclear palisading, whereas Antoni B tissues exhibit a considerable degree of cell pleomorphism in loosely arranged reticulum network. However, both of these tissues are usually found in most neurilemmomas, and there is probably no relation to frequency of local recurrence or malignant change<sup>12</sup>).

The advocated treatment of these tumors is sur-

gical extirpation. Since these tumors are clinically and histologically benign, preservation of the continuity of the neural pathway is emphasized while the tumor is being extirpated. Several methods have been described for preserving the neural pathway<sup>2)3)7)13)</sup>. With adequate surgical exposure via a transcervical approach, most of these tumors can be usually enucleated, and the trunk of the cervical vagus nerve may be preserved because individual fibers of the vagus nerve are splayed over the surface of the tumor within a discrete capsule but not in the mass itself<sup>2)3)7)13)</sup>. If an adequate plane cannot be found even under microsurgical technique, a portion of the adherent fascicles of the nerve splayed over the tumor may be excised with the main specimen without sacrificing the nerve trunk. If it is technically difficult to preserve the nerve trunk intact, and if the lesion is less than 2cm in length, it may be possible to resect the involved segment and do microsurgical reanastomosis of the cut ends<sup>2)7)</sup>. When a situation requires the sacrifice of more than 2cm in length, an interposition with a free nerve grafting using a sensory nerve such as the greater auricular can also be considered<sup>2)13)</sup>.

Following adequate surgical extirpation, these tumors rarely recur<sup>7)8)</sup>. In our 4 patients, the results of intracapsular enucleation were quite satisfactory to cure the patients, although we observed 2 cases of transient postoperative vocal cord palsy. Therefore, we do think that this kind of surgery should be considered, whenever possible, before a decision is made to sacrifice the nerve.

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