The Journal of Laryngology and Otology
May 1996, Vol. **110**, pp. 485–489

Parapharyngeal vagal neurilemmoma extending to the jugular foramen

Eiji Yumoto, M.D., Koshiro Nakamura, M.D., Toshihiro Mori, M.D., Naoaki Yanagihara, M.D.

Abstract
A large vagal neurilemmoma in a 33-year-old man is reported. He complained of slowly progressive palsy of the tongue on the left side. Weakness of soft palate movement was also noted. Magnetic resonance imaging (MRI) revealed a tumour in the left parapharyngeal space with partial extension to the posterior cranial fossa through the jugular foramen. Carotid angiography revealed avascularity of the tumour and anterior shift of the left internal carotid artery. The venous phase showed no blood flow in the internal jugular vein. The tumour was successfully extirpated via a transmandibular transpterygoid approach. Although vagus nerve dysfunction was not observed pre-operatively, the tumour was identified as a neurilemmoma arising from the vagus nerve. The surgical approach should be selected according to the lesion in individual patients. Since neurilemmoma is benign in nature, minimal post-operative sequelae should be expected.

Key words: Neurilemmoma; Vagus nerve

Introduction

The parapharyngeal space is anatomically complicated, and includes large blood vessels and cranial nerves in its poststyloid compartment. Tumours arising in the parapharyngeal space are rare and show a variety of histological characteristics. Among them, parotid gland tumours and neurogenic tumours are the most common (Work and Hybels, 1974; Som *et al.*, 1981; Stell *et al.*, 1985; Carrau *et al.*, 1990). We found a large vagal neurilemmoma in the parapharyngeal space which partially extended into the posterior cranial base through the jugular foramen. The tumour was totally removed via the transmandibular transpterygoid approach without any surgical sequelae. This article describes the clinical course and operative procedure in this patient.

Case report

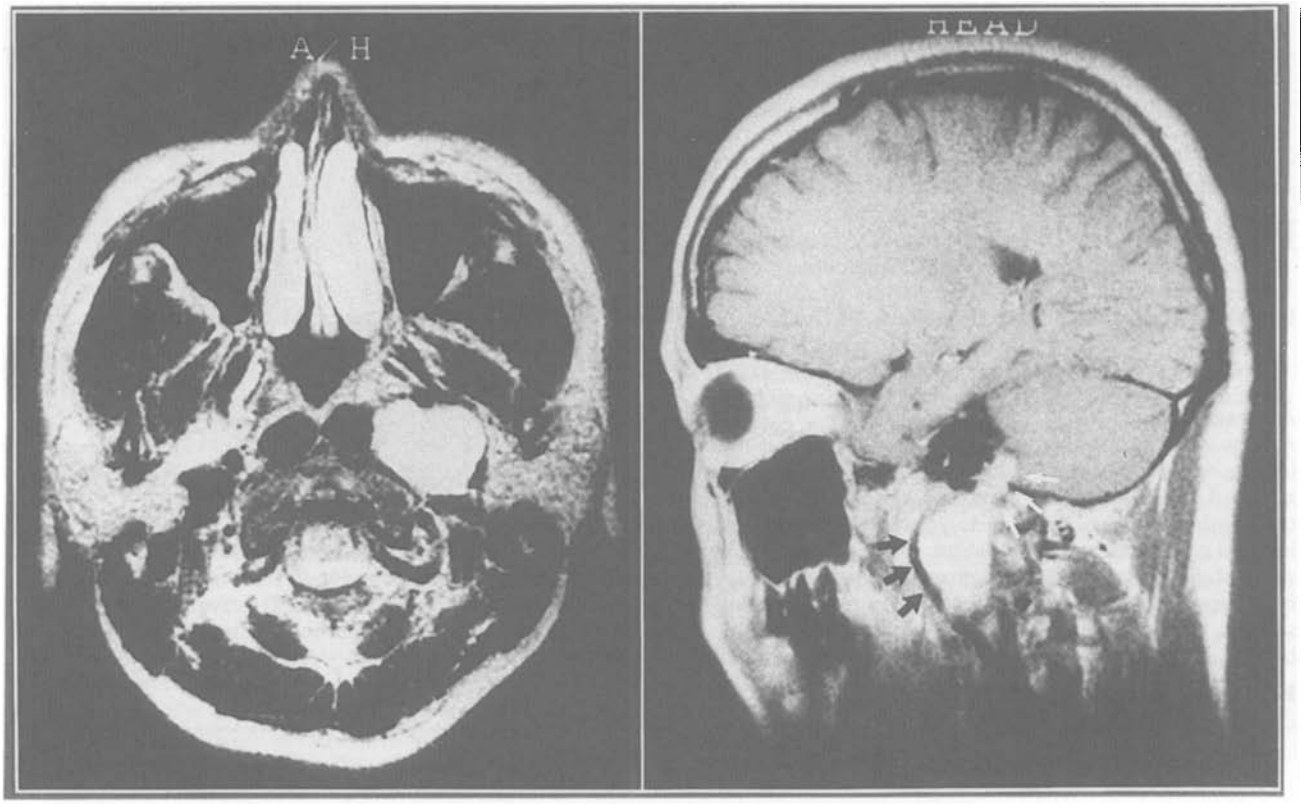
A 33-year-old man had noticed slowly progressive palsy of the tongue on the left side for a few years. He had consulted a local ENT clinic and was referred to us because a computed tomography (CT) examination had revealed a large tumour in the left parapharyngeal space. He did not report any other symptoms.

He was admitted to Ehime University Hospital on January 8, 1990. On admission, weakness in the elevation of the soft palate on the left side was noted. Movement of the vocal fold was not disturbed, and no cranial nerve dysfunction was observed except that of the glossopharyngeus and hypoglossal nerves on the left side. Palpation of the neck including the submandibular region and inspection of the oral cavity did not suggest the presence of a neoplastic lesion. Rigid nasal endoscopic examination revealed a local swelling in the area over the left tubal orifice and Rosenmuller fossa.

An MRI scan revealed a tumour (Figure 1) which extended to the posterior cranial fossa through the jugular foramen (Figure 1B, arrows). The left internal carotid artery (ICA) was displaced anteriorly (black arrows). Digital subtraction angiography confirmed the anterior shift of the left ICA and avascularity of the tumour (Figure 2A). The venous-phase left ICA angiography demonstrated no blood flow in the lateral sinus or internal jugular vein on the affected side (Figure 2B). Although vagus nerve dysfunction was not observed, a neurilemmoma arising from the vagus nerve was considered to be quite possible.

On January 23, 1990, the tumour was successfully resected through a transmandibular transpterygoid approach (Yumoto *et al.*, 1994). The first skin incision was made in parallel with the lower border of the mandible. Although the submandibular gland was removed, access to the lower border of the tumour could not be obtained. The skin incision was extended anteriorly to the midline, dividing the lower lip, and then posteriorly across the neck and superiorly to just below the auricular lobulus (Figure 3A). A mucosal incision was made from the corner of the lip incision, proceeding posteriorly along the lower gingivolabial sulcus (Figure 3B). Soft tissues including skin, the masseter muscle, and parotid gland were elevated from the mandible and retracted upward. The mandible was cut posterior to the third molar tooth. The coronoid process was resected to facilitate mobility of the ascending ramus of the mandible, which was then retracted upward. The exposed medial and lateral pterygoid muscles were divided (Figure 3C). The stylohyoid, stylopharyngeus, and digastric muscles were severed to expose the lateral surface of the tumour. The encapsulated

From the Department of Otolaryngology, School of Medicine, Ehime University, Shigenobu-cho, Onsen-gun, Ehime, 791-02, Japan.
Accepted for publication: 17 February 1996.

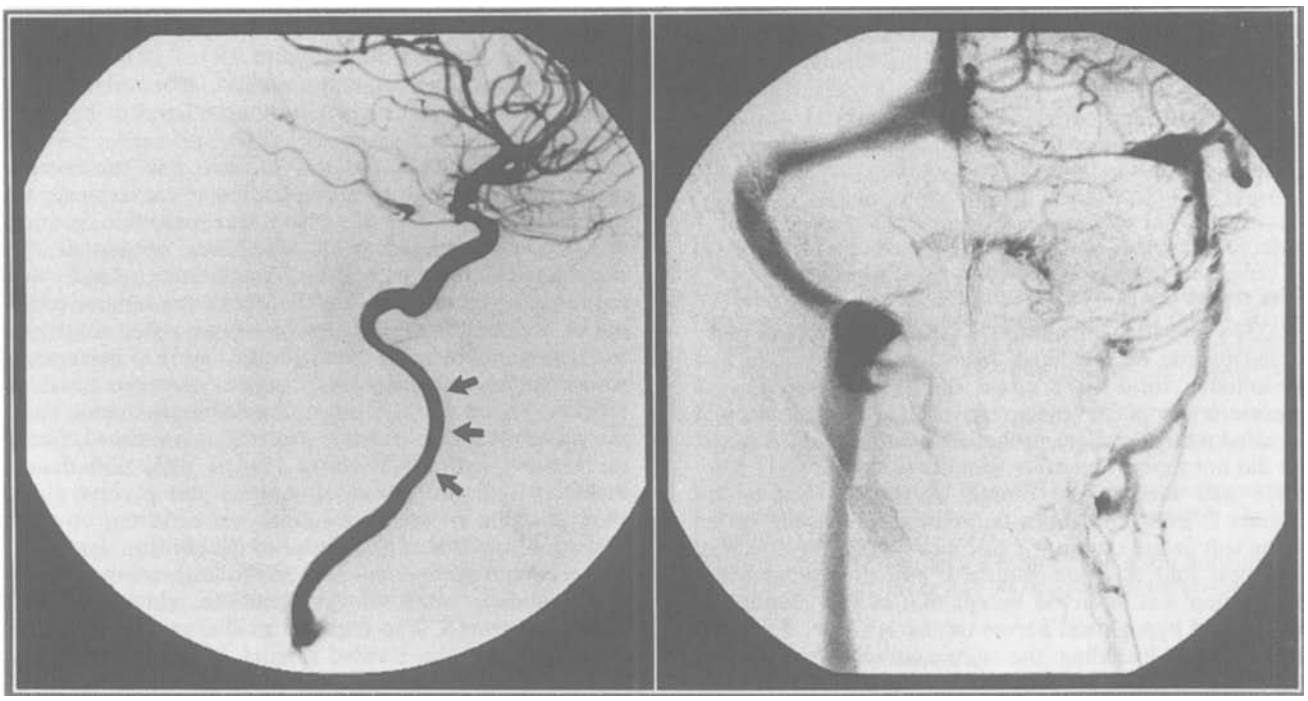


(A) (B)

FIG. 1

T_1-weighted magnetic resonance images with gadolinium enhancement. (A): Large parapharyngeal tumour. (B): Extension through the jugular foramen (white arrows) and the anteriorly shifted internal carotid artery (black arrows) are shown.

tumour was dissected from the laterally displaced external carotid artery. Intraoperative examination of a frozen section of a portion of the tumour revealed that it was a benign neurilemmoma. Multifocal bleeding from the pterygoid venous plexus was controlled by careful ligations. The hypoglossal nerve was found to be compressed by the tumour. The vagus nerve, identified at the level of the carotid bifurcation and followed upward, was firmly



(A) (B)

FIG. 2

Digital subtraction angiography. (A): The internal carotid artery (arrows) is shifted anterior to tumour. (B): Venous-phase angiography shows no blood flow in the lateral sinus or internal jugular vein on the affected side.

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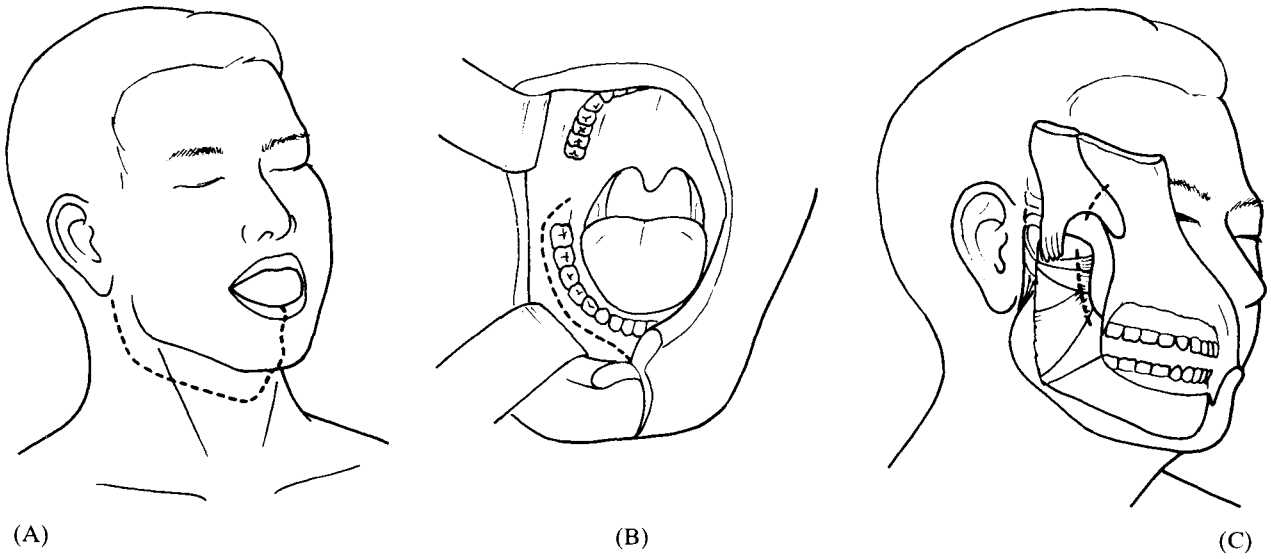

FIG. 3

Schematic drawing of the surgical procedure. (A): Skin incision. (B): Incision of the oral mucosa. (C): Approach to the parapharyngeal space by upward retraction of the ascending ramus of the mandible and soft tissues. The internal and external pterygoid muscles are severed.

connected with the tumour, suggesting that the tumour had originated from the vagus nerve. The tumour was carefully dissected toward the skull base, from the ICA anteriorly and from the internal jugular vein and hypoglossal nerve posteriorly. Finally, the tumour was extirpated en masse, and measured 55 × 33 × 20 mm (Figure 4). Leakage of cerebrospinal fluid from the jugular foramen was noted. Placement of a double layer of the fascia lata, sealed with bioactive adhesive and masses of fat, was useful in stopping the leak. The dead cavity left after removal of the tumour was packed with the pterygoid muscles. The retracted ascending ramus of the mandible assumed its original position and was fixed with a miniplate system. Tracheostomy was performed to maintain the airway during the immediate post-operative period. Finally, a drainage tube was inserted into the subarachnoid space through a lumbar puncture to prevent elevation of the intracranial pressure.

The drainage tube in the subarachnoid cavity was removed one week after the operation and the tracheostoma was closed three days later. Although the left recurrent and superior laryngeal nerves (RLN and SLN) were paralyzed post-operatively, the patient had no difficulty in deglutition or speech. Trismus seen in the immediate post-operative periods was restored to normal two months after the operation (Figure 5). The patient's post-operative course has been uneventful. There are no signs of recurrence at the time of this writing, four years and six months post-operatively. Unfortunately, the deficits of the IX, X and XII nerves persist. Since the patient has not complained about his moderately breathy voice, we did not perform phonosurgical treatment.

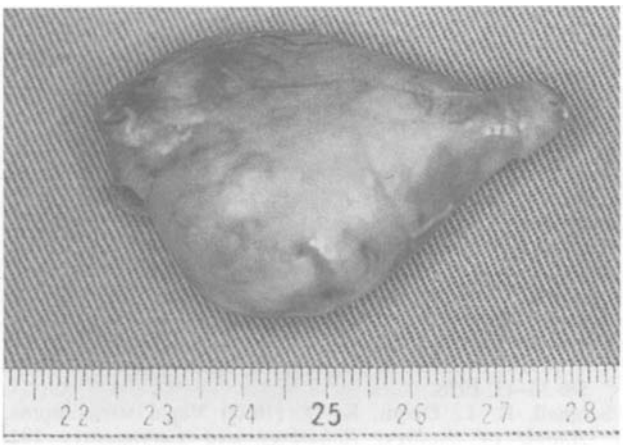

FIG. 4

Macroscopic appearance of the totally removed tumour. Scale is in centrimetres.

Discussion

The parapharyngeal space includes cranial nerves IX-XII, the sympathetic nerve trunk, ICA, and internal jugular vein, mostly in the poststyloid compartment.

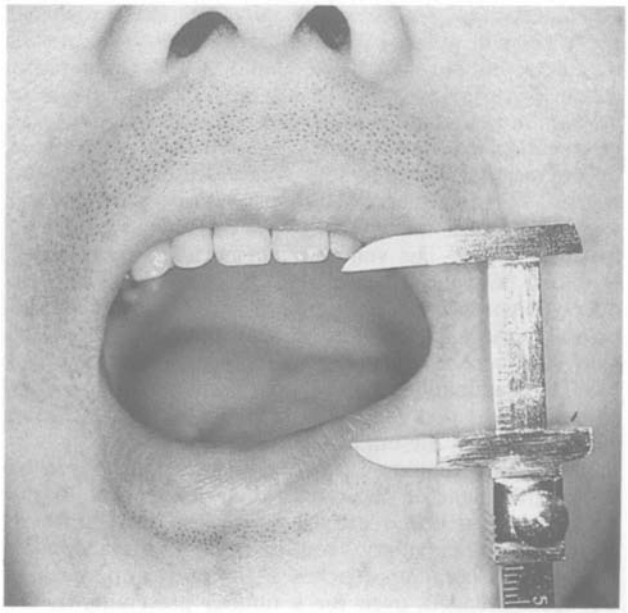

FIG. 5

Post-operative facial view, showing trismus measurement.

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Among a variety of histologically characteristic tumours arising in this space, neurilemmoma and neurofibroma have been reported to occur at incidences ranging from 9.3 per cent to 28 per cent according to the English literature (Work and Hybels, 1974; Som *et al.*, 1981; Stell *et al.*, 1985; Carrau *et al.*, 1990). In contrast, a relatively higher incidence of parapharyngeal neurilemmoma in Japan has been reported by Miyahara *et al.* (1978), who reviewed cases of parapharyngeal neoplasm reported in the Japanese literature and found that 42 (55 per cent) of the total of 77 tumours were neurilemmomas. Cranial nerves IX-XII and the sympathetic nerve trunk in the parapharyngeal space are encased by Schwann cells and may therefore develop neurilemmoma. According to Maniglia *et al.* (1979), approximately half of reported parapharyngeal neurilemmomas are considered to be of vagus nerve origin.

The parapharyngeal space is surrounded laterally by the mandible and deep lobe of the parotid gland, posteriorly by the vertebral column, and superiorly by the skull base. Thus, these bony walls usually allow the medial and inferior growth of tumours originating in this space, resulting in a mass in the upper neck and swelling in the tonsillar region. However, the tumour reported herein had expanded superiorly to the posterior cranial fossa through the jugular foramen. As Green *et al.* (1988) reported, neurilemmoma of the vagus nerve has a predilection to originate near the nodose ganglion, which is located very high in the neck just posterior to the ICA and inferior to the jugular foramen. Since a parapharyngeal neurilemmoma grows very slowly and does not induce any marked symptoms, in most cases it is not diagnosed until a neck mass or multiple cranial nerve paralysis appears. Crumley and Wilson (1984), Horn *et al.* (1985), and Franklin *et al.* (1989) reported cases in which neurilemmoma of the jugular foramen extended widely into the posterior cranial fossa.

A mass in the neck or throat is the most common complaint in parapharyngeal neurilemmoma. Cranial nerve deficits are uncommon except in patients with large tumours and are not always indicative of the nerve of origin. Work and Hybels (1974) and Schmall and Dolan (1992) reported cases of vagal neurilemmoma accompanied by hypoglossal nerve paralysis. Left RLN, glossopharyngeus nerve, and accessory nerve paralysis was also observed in the patient reported by Schmall and Dolan (1992). The present patient manifested left hypoglossal nerve palsy of gradual onset and glossopharyngeal nerve weakness, but no symptoms related to the vagus nerve deficit. Nevertheless, vagal neurilemmoma was thought to be the most probable diagnosis prior to the surgical intervention. The pre-operative diagnosis was made based on the location of the tumour demonstrated by MRI, the anterior shift of the left ICA clearly revealed by MRI and angiography, and the avascularity of the tumour confirmed by angiography. The venous-phase angiography revealed no blood flow in the lateral sinus or internal jugular vein due to compression of the tumour. These findings were also helpful in the selection of the operative route. Jacobs *et al.* (1987) retrospectively evaluated the usefulness of CT and MRI in examination of vagus neuropathy in 48 patients, and suggested that MRI is optimal for imaging a lesion located between the brain stem and oropharynx.

Since neurilemmoma is radioresistant, surgical resection is the treatment of choice. The surgical routes to the parapharyngeal space are classified into the transoral, transcervicosubmaxillary, transparotid, transmandibular, and infratemporal approaches. A transoral approach is not recommended, even for a tumour presenting in the oropharynx because of the possibilities of vascular or neural injury and a restricted operative field. In a patient with a tumour confined to the parapharyngeal space, the surgical approach is selected based on the size and location of the tumour and its relation to the large vessels. When a tumour extends to the posterior cranial fossa, a suboccipital craniotomy is usually also made either simultaneously or separately (Arenberg and McCreary, 1971; Crumley and Wilson, 1984). Horn *et al.* (1985) applied the translabyrinthine, retrolabyrinthine, and infratemporal approaches in the resection of jugular foramen neurilemmomas showing varying degree and direction of tumour extension.

Franklin *et al.* (1989) utilized the infratemporal approach for the removal of jugular foramen neurilemmomas in seven patients, all of whom had advanced lesions classified as C_2 or C_3, indicating tumour involvement of the ICA above the carotid foramen. Their method led to loss of hearing in every patient. Furthermore, with this method it is difficult to dissect the medial and inferior surfaces of the tumour under direct vision, and the distance to the operative field from the surgeon's hand is not shorter by this route. The lesion in the present patient was classified as C_1, indicating that the tumour filled the jugular foramen with resultant bone expansion and that the ICA was not involved above the carotid foramen. We severed the mandible posterior to the third molar tooth, not at the midline, and retracted the ascending ramus and soft tissues superiorly. This transmandibular transpterygoid approach provides sufficient exposure of the tumour, great vessels, and cranial nerves. It also offers a relatively shorter route to the operative field. Thus, a lesion without involvement of the ICA above the carotid foramen can be managed by this approach. In addition, hearing and facial nerve function are not sacrificed. The surgical approach should be selected according to the lesion in each patient to minimize post-operative sequelae. Since neurilemmoma is benign in nature, every effort should be made to preserve important nerve functions.

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Address for correspondence:
Eiji Yumoto, M.D.,
Department of Otolaryngology,
School of Medicine, Ehime University,
Shigenobu-cho, Onsen-gun,
Ehime 791-02, Japan.

Fax: 81-899-64-4613

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