"PARAPHARYNGEAL" NEUROGENIC TUMOURS

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It is uncommon to find tumours of neurogenic origin in the neck, and they are even more rare in the parapharyngeal space. Despite this, certain types of tumour seem to have a special predilection for settling in the neck. It is, in fact, known that about 20 per cent. of all neurinomas occur precisely at this site. Those types of tumour which are met with are the benign and malignant neurinoma, ganglioneurinoma, sympathicoblastoma and paraganglioma. The most usual site of origin is the sympathetic chain (Amano, 1934; Brandt, 1953; Conley, 1955; Cranmer, 1958; Hanley and Davol, 1952; Huet, 1952; Lindén and Hamberger, 1954; Terracol and Causse, 1959), but tumours originating from the cranial nerves have also been encountered. Thus, Cohn (1928) reported a case in which the tumour arose in the accessory nerve, Sekiguchi and Oije (1926) described neurinomas taking their origin in the vagus nerve, and Erlich and Martin (1943) those originating in the phrenic nerve. Petit-Dutaillis *et al.* (1958) gave an account of multiple neurinomas arising in the hypoglossal and vagus nerves. According to Terracol and Causse (1959) a neurinoma of the glossopharyngeal or hypoglossal nerve is exceedingly rare.

Anatomy

To understand the clinical features and therapy of these tumours, it is necessary to be acquainted with the anatomy of the region where they are found. The space between the spinal column and the visceral cranium is denoted as the craniovertebral space. In this region, the styloid process is perhaps the most important keystone for the surgeon. This is because it is the site of origin of the stylopharyngeal septum, which is a musculofibrous sail, running between the styloid process and the lateral pharyngeal wall (Fig. 1). This septum divides the *craniovertebral space* into two parts; one anterior, which is known as the *parapharyngeal space* and one posterior, denoted as the *retropharyngeal space*. In its lateral border, the septum is strengthened by the styloglossal and stylopharyngeal muscles and the stylomandibular ligament. The stylopharyngeal septum serves as a supporting plate for those nerves and vessels running toward the pharynx. The vessels running in this direction are usually given off from the external

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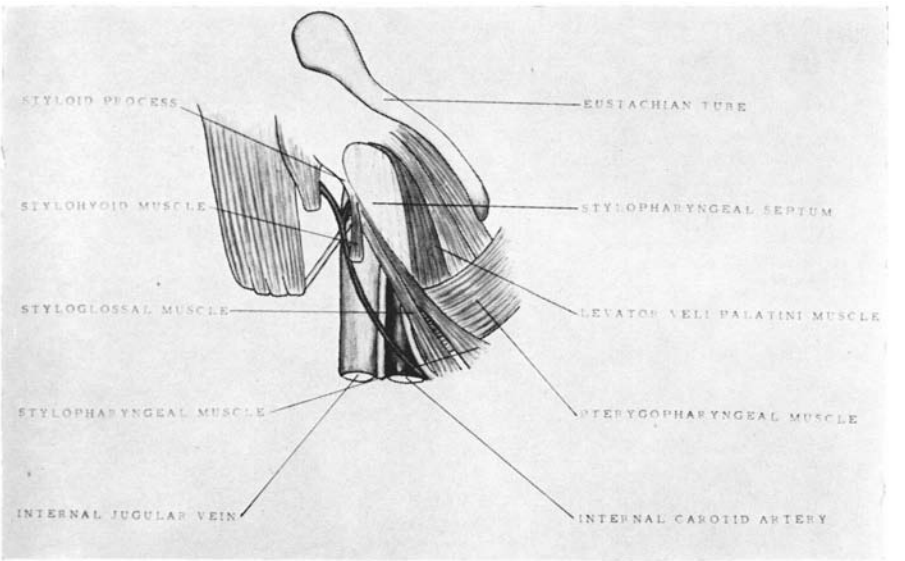


FIG. 1.
The styloid process as the keystone.

carotid artery. Only in rare cases does the ascending pharyngeal artery arise from the internal carotid artery, after which it follows the septum in the medial direction.

Medial to the styloid process, and hidden behind the fairly thick septum, the internal carotid artery ascends toward the skull. Behind the septum is also the opening of the jugular foramen, with the internal jugular vein. Initially, it has no contact with the septum, but further caudally it touches on the muscles given off from the styloid process. In addition, the anterior part of the jugular foramen is crossed by three nerves coming from the skull, namely, the glossopharyngeal, vagus and accessory nerves. The hypoglossal nerve from the hypoglossal canal also enters from behind the vein. At their exit from the skull, all four nerves lie behind the internal carotid artery, whereas laterally they run in different directions. Thus, the glossopharyngeal nerve passes downward, lateral to the internal carotid artery, to the inferior border of the stylopharyngeal muscle, after which it accompanies the stylopharyngeal septum to the tongue and throat.

The only one of the nerves which, during its whole course downward, follows the sheath of the vessels is the vagus nerve, which lies between the artery and the vein (Fig. 2). Via the stylopharyngeal septum, it sends small branches to the throat. High up in the retro-pharyngeal space, it also gives off the superior laryngeal nerve which runs, behind the internal carotid artery, forward and downward toward the larynx.

The accessory nerve runs laterally from the jugular foramen, in front

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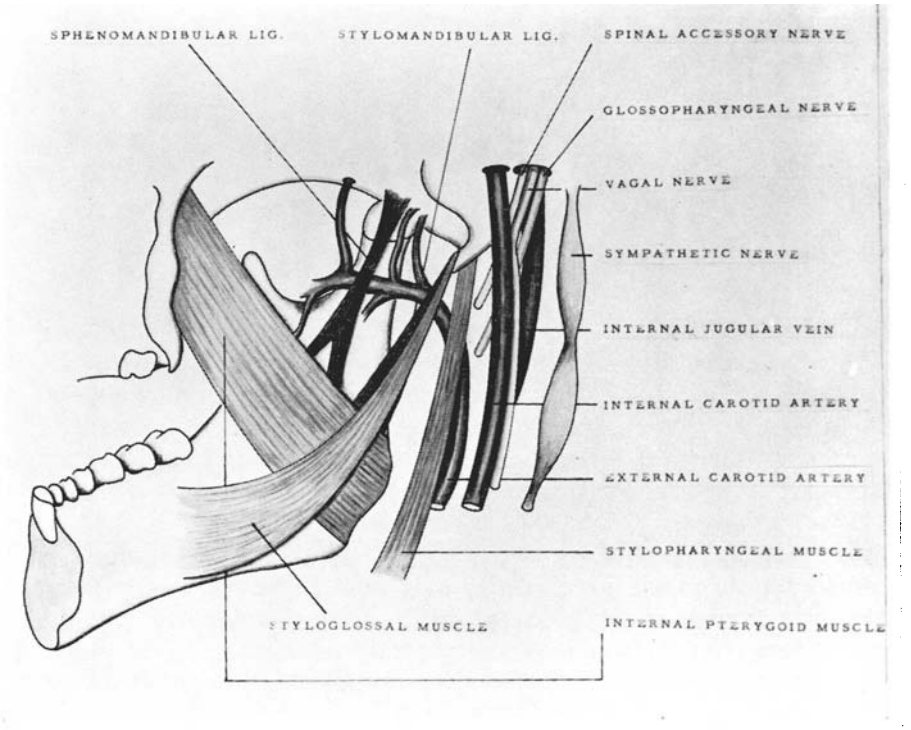


FIG. 2.
Vagus nerve relationships.

of the internal jugular vein, and emerges—below the styloid process—between the digastric and sternocleidomastoid muscles, after which it passes downward toward the trapezius muscle.

Close to its exit from the hypoglossal canal, the hypoglossal nerve crosses the posterior part of the internal carotid artery, and passes forward in the direction of the tongue, in a superficial arch lying on the arterial branches.

Finally, the sympathetic chain extends from the base of the skull in the caudal direction, situated in front of the prevertebral fascia but all the time behind the internal carotid artery.

A common localization of the neurogenic tumours is precisely the retro-pharyngeal space, and the tumour is often so large that it presents as a lump not only inside the throat, but also outside it. The subsequent extension of the tumour is, naturally, determined by its site of origin. Thus, a tumour originating in the sympathetic chain is primarily situated behind the internal carotid artery, but later usually tries to expand on the medial side of the vessel, and then gradually continues to grow behind the stylopharyngeal septum, toward the throat (Fig. 3). Here, it presses the

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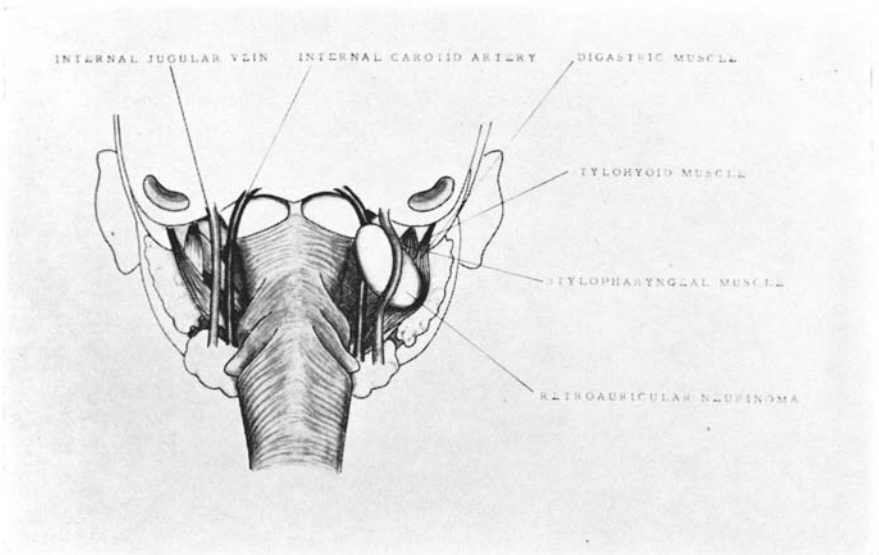


FIG. 3.
Retro-auricular neurinoma.

posterior part of the pharyngeal wall behind the tonsillar region and above it. It is not until the tumour has reached the size of an orange that it penetrates below the inferior border of the styloglossus and stylopharyngeal muscles, and can then be palpated on the outside of the neck.

Sometimes, however, the tumour may also expand on the lateral side of the internal carotid artery, specially if it arises in any one of the last four cerebral nerves (Fig. 4). It then extends laterally between the artery and the vein, and when it reaches the styloid process it is diverted in the posterior direction toward the narrow space between the mastoid process and the spinal column. In this case, the tumour is located below and behind the insertion of the sternocleidomastoid muscle. Consequently, the great vessels lie medial to the tumour, and are displaced inward toward the pharynx, where on bimanual palpation the pulsations can sometimes be felt more distinctly than usual.

Symptoms and Signs

Initially, the neurogenic tumours seldom produce any noteworthy symptoms or signs. It is not until they reach a certain size that they may produce pressure symptoms. Sometimes the patient can have a feeling of pressure or of a foreign body on one side of the neck or in the throat. If the tumour presses upward toward the base of the skull it can, by compression of the Eustachian tube, produce a feeling of increasing deafness

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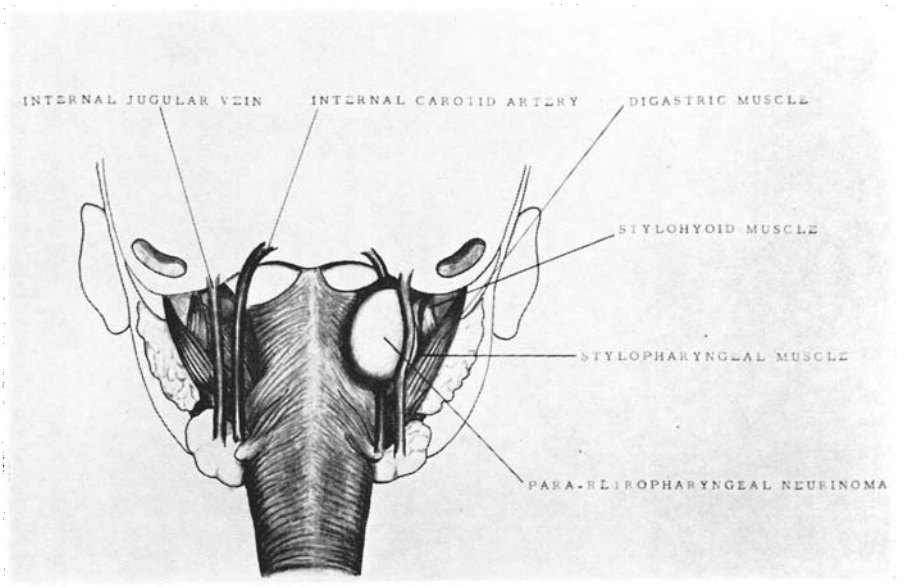


FIG. 4.
Para-retropharyngeal neurinoma.

on the side involved. Although the nerve in which the tumour arises often shows no signs of involvement, this occasionally happens. It is therefore of some diagnostic value to be aware of the nerves from which symptoms are to be regarded as suspicious.

A tumour originating in the *sympathetic nerve* often produces no symptoms from this nerve, but a Horner's syndrome may appear. Lindén and Hamberger (1954) have demonstrated an additional sign of loss from the sympathetic system, namely, hyperæmia and hyperplasia of the nasal mucosa on the same side as the tumour.

A tumour arising in the *glossopharyngeal nerve* may be situated in the jugular foramen, and then produces largely the same symptoms as a jugular tumour. It can give sensory disturbances in the throat, and loss of taste on the posterior third of the tongue. Furthermore, since this nerve supplies the stylopharyngeal muscle, damage to it decreases the capacity to elevate the throat during the act of swallowing. This produces the "signe de rideau" toward the healthy side.

A tumour of the *vagus nerve* may occasionally be associated with damage or disturbances of various organs. Thus, there may be a loss of sensation in the pharynx and the inlet of the larynx, so that the patient is apt to "swallow the wrong way". In addition, unilateral paralysis of the larynx occurs, as well as cardiac symptoms.

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"Parapharyngeal" Neurogenic Tumours

A neurinoma of the *accessory nerve* may produce motor disturbances of the trapezius muscle, with difficulty in raising the arm above the shoulder plane.

In the rare cases of a tumour of the *hypoglossal nerve*, motor disturbances of the tongue may appear.

Diagnosis

Every patient with diffuse or unclear complaints in the region of the neck should undergo bimanual palpation, with one finger in the throat and another on the outside of the neck below the mandibular angle. For the experienced examiner, it is easy to palpate through the whole retropharyngeal space.

If a tumour penetrates toward the lateral wall of the pharynx, its extension can sometimes give a certain hint, and the presence of any neurological symptoms may also give an idea of the location. Since the neurogenic tumour extends from the parastyloid recess behind the stylopharyngeal septum, it reaches the pharynx on the borderline of the retropharyngeal space. This implies that the lateral, posterior part of the pharyngeal space is pushed inward, and the tonsil is displaced downward and forward. Consequently, the lump will be localized to the posterior palatine arch, in contrast to the swelling found in the presence of a parapharyngeal parotid tumour, which displaces the tonsil in the direct medial direction, and possibly somewhat downward.

In tumours extending toward the lateral wall of the pharynx, the mucous membrane is always found to be easily movable against the tumour. This is because the latter is located outside the constrictor pharyngis muscle and the pharyngeal fascia.

Carotid angiography and retrograde jugular angiography are valuable tools for establishing the location of the tumour. They show whether the tumour is situated medial to the internal carotid artery, or whether it extends in the lateral or caudal direction. A malignant tumour sometimes infiltrates these vessels, which can perhaps be disclosed by angiography. In addition, a roentgenologic examination of the skull should be made, to determine whether the bones at its base have been invaded.

The histological diagnosis is established by puncture or biopsy. The question then arises of where the specimen should be taken. A tumour situated outside the constrictor pharyngis superior muscle may, after biopsy through the pharynx, become adherent to the mucosa and musculature, and therefore present difficulties at subsequent operation by the external approach. Consequently, if the specimen is taken from the pharynx, it should not be excised, but one should confine oneself to puncture, which generally suffices for a diagnosis. For puncture, it is naturally also possible to enter directly behind the submandibular gland,

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but it may then be difficult to locate the tumour with the puncture needle, and to obtain a representative specimen.

Differential Diagnosis

In addition to the neurogenic tumours, those most commonly found in the true parapharyngeal space are tumours of the parotid gland. But other types of tumours can also be seen in this region. Thus, reports are found of *fibrolipoma* (Howarth, 1930), *lymphoma* (New and Childrey, 1931) and *lymphatic leukemia* confined to the parotid glands (Patey and Thackray, 1957). An aneurysm of the internal carotid artery is also found; it is manifested in the pharynx in the form of expansile pulsations. Another vascular disease is a tumour of the *carotid body* which, although it is generally visible on the outside of the neck, may occasionally press the lateral wall of the pharynx inward (Morfit, 1955).

Other lesions that should be borne in mind are *chordoma* (André and Laccourreye, 1962), *congenital cysts* arising in the first branchial arch (Ombrédanne and Goddé, 1962) and the possibility that a *cholesteatoma* may empty into the parapharyngeal space through the lower border of the pars petrosa (Goddé, 1962).

Therapy

Because of their location, the neurogenic tumours have often wrongly been regarded as inoperable. In view of the proximity of the tumour to the great vessels of the neck, the bleeding that may occur can be overcome only with the use of an external approach. Planning of the operation is determined by the type and location of the tumour. One of the problems of operation in this region is the lack of space. Fairly good access to the operative field can be achieved by hyperextending the patient's head and turning it to the side. Further improvement is obtained if an assistant keeps the lower jaw subluxated forward with a retractor, which increases the space by almost 50 per cent. In extremely inaccessible cases, the space can be further widened by dividing the mandible. When the tumour grows in the posterior direction on the inside of the mastoid process, it may be necessary—to obtain a sufficient view—to divide the mastoid process, and then push the sternocleidomastoid muscle downward (Petit-Dutaillis *et al.*, 1958).

Operation is started by making an incision from the region of the mastoid process, downward and caudally in an arch about 4 cm. from the mandibular angle, as far as the chin. The skin, including the platysma, is dissected free from the underlying muscles, and the flap tied upward in one of the operation cloths. The bifurcation of the common carotid artery is then explored. The external carotid is ligated and divided. The proximal stump is then lifted toward the mandible, which implies that all its

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branches, as well as the stylopharyngeal septum, will be lifted up toward the internal pterygoid muscle. To obtain additional space, it is sometimes advisable to resect the retromandibular process of the parotid gland. To reach the region behind this septum more easily, the styloid process is dissected free, as well as the facial nerve, after which the styloid process is fractured forward, so that the styloglossus and stylopharyngeal muscles can relax. It is seldom necessary to divide the digastric and stylohyoid muscles. The internal carotid artery, vagus nerve and internal jugular vein are then dissected free from the sympathetic chain, and retracted with a hook. This procedure creates a fairly large space in front of the spinal column, which allows visual inspection of the operative field.

To avoid a fall in blood pressure on traction of the vagus nerve, the conduction of the nerve should be blocked by local anæsthesia. It is then possible, under visual control, to excise the tumour, as well as to inspect the nerve from which it arises and, if necessary, also to suture the nerve.

In the presence of a tumour growing laterally behind the styloid process, it is advisable to cut this process and to displace the whole stylopharyngeal septum medially and forward. By cautious dissection, the tumour can then be freed from the large vessels, which are situated on its medial side, after which it is carefully pushed caudally and removed. If the vein is ruptured, it is easy to suture or to tamponade, if the rupture should be situated too close to the jugular foramen. In rare cases one can, as stated earlier, be forced to divide the mastoid process to obtain satisfactory access to the tumour.

In the presence of a *malignant tumour* of the parapharyngeal space, it is often impossible to perform a completely radical operation, because the tumour very rapidly penetrates the fascia and muscles, and then infiltrates the large vessels. When the tumour is suspected to be malignant, it is recommended always to place a loop around the common carotid artery prophylactically, to avoid profuse hæmorrhage if the internal carotid has been injured during dissection. Post-operative irradiation therapy should be considered.

Prognosis

The prognosis is considered to be good in all forms of benign neurogenic "parapharyngeal" tumours. The functional prognosis is also very good, even in the cases where both nerve graft and suture of vessels have been required (Conley, 1955).

Summary

Neurogenic "parapharyngeal" tumours are almost always located behind the stylopharyngeal septum, and most frequently arise from the sympathetic chain, but occasionally from other cranial nerves. An account

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is given of the clinical features and diagnosis of these tumours. The most suitable process for their removal is described. Using the external approach, the tumour can be excised, under visual control, without any great difficulty. A report is given of three cases.

Case Reports

Case I, a 40-year-old woman. She had a history of multiple neurinomas since childhood. For the past few years, she had a lump in the right mandibular angle, which had started to grow more rapidly in the last 6 months. The tumour produced no distress. Examination disclosed a somewhat painful resistance, the size of a walnut, below the right mandibular angle. It also filled the right side of the throat, and extended downward to the aryepiglottic region. Roentgenologic examination of the cervical spine showed a considerable increase in diameter of the intervertebral foramen on both sides.

Operation revealed a tumour, 6 × 3 cm., in the parastyloid recess, extending inward toward the pharynx. Closer to the spinal column, another tumour of almost the same size was present, and was also removed. There was no erosion of the spinal column. *Histological diagnosis:* neurofibroma. Postoperatively, the patient was asymptomatic.

Case II, a 53-year-old woman. She had previously been given irradiation therapy for a lymphoma of the uterus. For the past 6 months she had pain in the left arm, radiating into the back of the neck on the left side. Examination showed a lump, the size of a thumb tip, below the insertion of the sternocleidomastoid muscle. Angiography of the jugular vein disclosed marked compression of the vein from the lateral side; thus, in the sagittal plane, it was only 3 mm. wide. Roentgenologic examination of the spinal column and base of the skull showed no pathological changes.

At operation, a tumour 3 × 4 cm. was found; it lay lateral to the internal jugular vein, and extended as far as the base of the skull. The tumour was removed *in toto*. *Histological diagnosis:* neurinoma. Postoperatively, a Horner's syndrome appeared.

Case III, a 49-year-old man. Two years previously, he had sought medical advice for difficulty in swallowing and a sensation of a lump in his throat, as well as a slight cough. Examination showed right-sided paralysis of the larynx and pharynx, with a "signe de rideau" to the left. There was atrophy of the right side of the tongue. At that time, no tumour was demonstrable, and the patient was referred to a neurological clinic. Two years later, increasing deafness appeared on the right side, and pain in the right side of the neck toward the ear. Examination disclosed a tumour bulging behind the right tonsillar fossa, extending from the base of the skull fairly far downward in the throat.

Operation revealed a tumour in the para-retropharyngeal space, slightly larger than a walnut, extending to the base of the skull. It was removed *in toto*. *Histological diagnosis:* neurofibrosarcoma. Postoperative irradiation therapy was considered, but was not regarded to be immediately necessary.

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REFERENCES

AMANO, M. (1934) *Zbl. Chir.*, **61**, 78.
ANDRÉ, P., and LACCOURREYE (1962) *Ann. Oto-laryng.* (*Paris*), **79**, 312.
BRANDT, U. (1953) *Acta oto-laryng.* (*Stockh.*), **43**, 503
COHN, I. (1928) *Arch. Surg.*, **17**, 117.
CONLEY, J. J. (1955) *Arch. Otolaryng.*, **61**, 167.
CRANMER, L. R. (1958) *Ann. Otol.* (*St. Louis*), **67**, 178.
ERLICH, H. E., and MARTIN, H. (1943) *Surg. Gynec. Obstet.*, **76**, 577.
FIGI, F. A., and STARK, D. B. (1953) *Laryngoscope* (*St. Louis*), **63**, 652.
GODDÉ, C. (1962) *Ann. Oto-laryng.* (*Paris*), **79**, 619.
HANLEY, J. S., and DAVOL, R. T. (1952) *Arch. Otolaryng.*, **56**, 207.
HOWARTH, W. (1930) *J. Laryng.*, **45**, 673.
HUET, P. C. (1952) *Ann. Oto-laryng.* (*Paris*), **69**, 519.
LINDÉN, A., and HAMBERGER, C. A. (1954) *Acta oto-laryng.* (*Stockh.*), Suppl. **116**, p. 205.
MORFIT, H. M. (1955) *Arch. Surg.*, **70**, 906.
NEW, G. B., and CHILDREY, J. H. (1931) *Arch. Otolaryng.*, **14**, 596.
OMBRÉDANNE, M., and GODDÉ, C. (1962) *Ann. Oto-laryng.* (*Paris*), **79**, 613.
PATEY, D. H., and THACKRAY, A. C. (1957) *Brit. J. Surg.*, **44**, 352.
PETIT-DUTAILLIS, D., CHAVANY, J. A., FISCHGOLD, H., PERTUISET, B., and BERDET, H. (1958) *Sem. Hôp. Paris*, **34**, 2389.
SEKIGUCHI, S., and OIJE, T. (1926) *Langenbecks Arch. klin. Chir.*, **143**, 113.
TERRACOL, J., and CAUSSE, J. (1959) *Ann. Chir.*, **13**, 1307.

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