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GLOSSO-PHARYNGEAL NEURALGIA*

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A COMPLETE diagnosis in diseases of the nervous system aims at establishing both the *site* of a lesion and its *pathology*; it must answer the questions, Where is the trouble? and What is its nature? In most instances a correct interpretation of the symptoms and signs presented by the patient will answer both questions. Often, however, the evidence suffices to elucidate only one aspect of the diagnostic problem. We can, for example, conclude from a characteristically Jacksonian fit which begins in the foot that there is a focal irritative lesion in the upper part of the opposite Rolandic cortex, but whether the cause be a neoplasm, an aneurysm or a localized patch of meningitis, we cannot tell. More rarely, the pathology of a lesion is known but its site eludes us. We know, for example, that Argyll-Robertson pupils point almost invariably to a syphilitic infection of the nervous system but the exact site of the lesion remains unknown. In chronic epidemic encephalitis, involuntary movements of the limbs and respiratory disorders have no definitely determined anatomical basis. If this point be appreciated we shall understand more clearly the significance of *neuralgia* or *nerve pain*.

Since irritation of the pain fibres in a sensory nerve at any point in their course may cause pain to be referred to the peripheral distribution of that nerve, we can anticipate finding neuralgias associated with any sensory spinal nerve, and with the Vth (trigeminal) and IXth (glosso-pharyngeal) cranial

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nerves, which are largely sensory. In rare instances the VIIth (geniculate herpes) and the Xth (superior laryngeal neuralgia) cranial nerves may be the sites of neuralgic discomforts.

If the patient describes the site of a pain accurately and the observer has a detailed knowledge of the anatomy of the nervous system, there should be no difficulty in recognizing " neuralgias " ; they correspond to the distribution of sensory nerves. But when we investigate these pains from the etiological standpoint we find that they are divisible into two groups : (1) those in which a gross pathological lesion involves the nerve, for example, a tumour, inflammatory exudate, hæmorrhage, neuritis, etc., and (2) those which occur in the absence of any demonstrable pathology even when the patient is observed over a long period of time.

Some authors use the term neuralgia for the latter group only. This leads to inevitable confusion. It is preferable to refer to both groups as *neuralgias*—classifying the former as *secondary neuralgias*, thus emphasizing that gross pathology is present which careful investigation should unmask ; and the latter as *primary neuralgias*. I doubt if it would be justifiable to separate these two groups on the basis of causation only. The differentiation is, however, amply justified because the two groups present clinical pictures which differ essentially from one another and each can, in most cases, be recognized solely by its clinical signs and symptoms.

The main clinical characteristics of the *primary neuralgias* are : (1) the pain is paroxysmal, occurring as stabbing, shock-like thrusts, excruciatingly severe, but of short duration lasting only seconds, and separated by intervals of complete freedom from pain, though of profound apprehension of its return ; (2) stimulation of certain defined points known as " trigger zones " will precipitate paroxysms of pain ; (3) there is an absence of any clinical evidence of loss of function of the nerve ; there is no objective sensory disturbance, no motor or secretory change except those which may be reflexly produced during the paroxysm ; occasionally trophic changes occur but it is doubtful if these result directly from neurotrophic influence ; (4) the pain recurs at intervals over a period of many years without there being any evidence of spread to neighbouring nerves ; and (5) the onset is usually in middle or old age.

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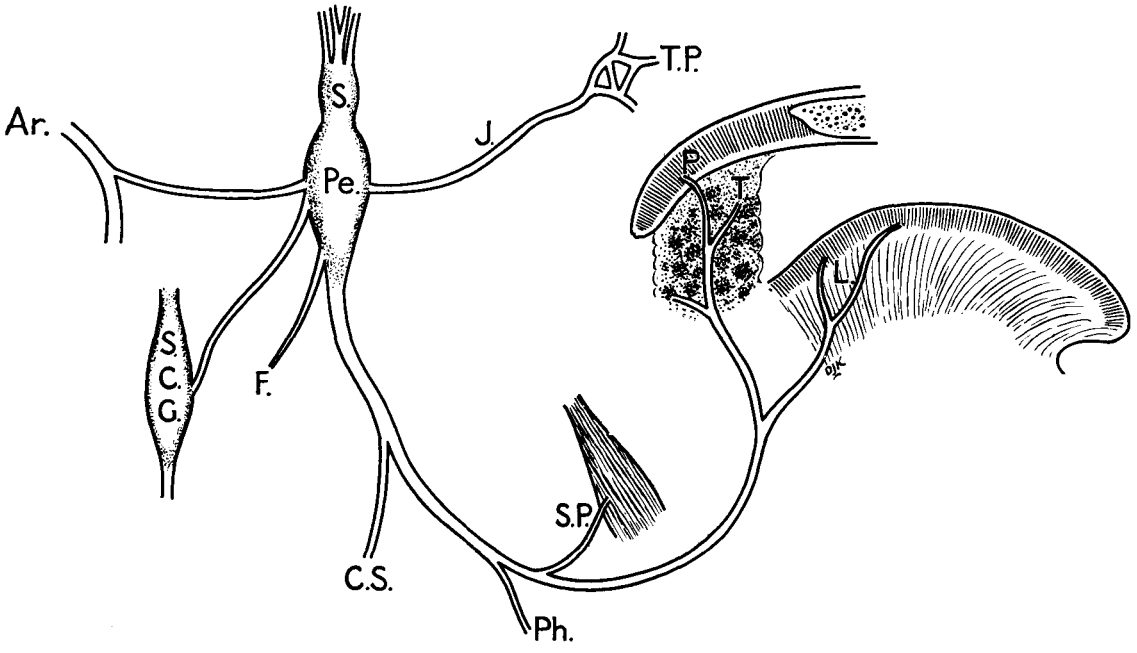


FIG. 1.

S.—Superior or jugular ganglion. Pe.—Petrous ganglion. Ar.—Arnold's nerve. S.C.G.—Superior cervical sympathetic ganglion. F.—Communicating branch to facial nerve. J.—Jacobsen's nerve. T.P.—Tympanic plexus. C.S.—Carotid Sinus nerve. Ph.—Branch to pharyngeal plexus. S.P.—Branch to stylopharyngeus muscle. L.—Lingual branches. T.—Tonsillar branches. P.—Palatal branches.

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Glosso-Pharyngeal Neuralgia

Secondary neuralgias are characterized by (1) a pain of a boring, aching type which is constantly present, though subject to exacerbations; (2) the pain is accompanied by objective evidence of nerve involvement, e.g. anæsthesia, reflex change, muscle wasting, etc.; (3) evidence of progressive involvement of neighbouring structures tends to appear; and (4) no age is immune.

The commonest site of primary neuralgia is the distribution of the trigeminal nerve (the so-called tic douloureux) and here the differentiation of our two neuralgic types is relatively simple. Secondary neuralgias of this nerve are extremely rare, but their presence can usually be demonstrated with certainty because the physical signs of direct pressure on, or irritation of, the Vth nerve are easily elicited. *Primary* and *secondary neuralgias* in the territory of the glosso-pharyngeal nerve are differentiated with greater difficulty because objective disturbances of this nerve's functions are not easy to demonstrate.

Fig. 1 shows diagrammatically the distribution of the glosso-pharyngeal nerve. It contains motor, sensory and secretory fibres. The motor fibres supply the stylo-pharyngeus muscle; the sensory convey sensation—not only from a part of the pharynx, palatine tonsil and the posterior third of the tongue, but also, as Ramsey Hunt has shown and as clinical evidence confirms in many instances, from the posterior part of the tympanum, the posterior wall of the auditory canal, part of the concha, a strip on the postero-medial surface of the auricle and the immediately adjacent mastoid region. It supplies secretory fibres to the parotid gland. The nerve arises from the brain by five or six fine radicals in the groove between the olive and restiform body in series with the roots of the vagus below. It passes outwards through the jugular foramen together with the vagus and spinal accessory nerves but clothed in a separate sheath of dura mater. It curves downwards and forwards to the interval between the hyoid bone and lower jaw, lying at first between the internal carotid artery and the internal jugular vein; and then between the internal and external carotid arteries to the pharynx. It passes round the stylo-pharyngeus muscle and the stylo-hyoid ligament and disappears beneath the hyoglossus muscle to its terminations in the tongue, tonsils and palate. Its branches and connections are shown in Fig. 1. The

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sensory loss following intracranial section of the left glosso-pharyngeal nerve is shown in Figs. 2 and 3. Taste is lost over the posterior third of the tongue.

The occurrence of glosso-pharyngeal neuralgias, both primary and secondary, is evidenced by the following brief notes of cases which have been under my care.

CASE I.—Mr. W. H., R.I. 50508. Aged 64 when first seen in July, 1931. Fourteen years previously he had begun to suffer from a pricking sensation in the right tonsillar and pharyngeal area in recurring attacks at intervals of some months lasting two to three weeks. A year or two after the onset his tonsils were enucleated without any appreciable effect on his discomfort. Eighteen months prior to his admission to hospital he experienced sudden intense pain in the right tonsillar area " as if his throat had been pierced with a red hot dagger " ; the pain was so severe that the sudden muscle jerking of the whole trunk which accompanied it flung him to the ground or out of bed ; this was regarded as hysterical. When I saw him he referred the pain to the right tonsillar and pharyngeal area, deep in the right ear and deep behind the angle of the jaw. Although it not infrequently appeared spontaneously, it could be excited by swallowing or touching the right tonsillar fossa, whilst anæsthetizing the surface of the pharynx and tonsillar area with cocaine prevented this " trigger " effect.

Careful examination of the nervous system proved negative. X-ray examination of the skull showed no bone change and a careful naso-pharyngeal overhaul revealed no abnormality. A diagnosis of primary glosso-pharyngeal neuralgia was made and on July 4th, 1931, Mr. Woolfenden performed *extra-cranial* avulsion of the glosso-pharyngeal nerve. This cured the neuralgia though it produced only slight objective sensory changes.

The patient remained perfectly free from pain and all discomfort until September, 1935, when the neuralgia recurred and was precipitated not only by swallowing but by talking and coughing. There was now no anæsthesia of the tongue and no loss of taste ; no motor change in the palate or pharyngeal wall was found and there was no evidence of disease elsewhere. He was examined by Mr. John Roberts who found a few small lymph granules on the right side of the posterior pharyngeal wall which were cauterized with temporary improvement of the pain ; but soon intensely severe paroxysms recurred and on March 11th, 1936, Mr. Woolfenden divided *intra-cranially* the root of the IXth nerve by Dandy's technique. There was a little post-operative intra-cranial hæmorrhage but after ten days the patient made a rapid recovery and was discharged three weeks after the operation quite fit. There has

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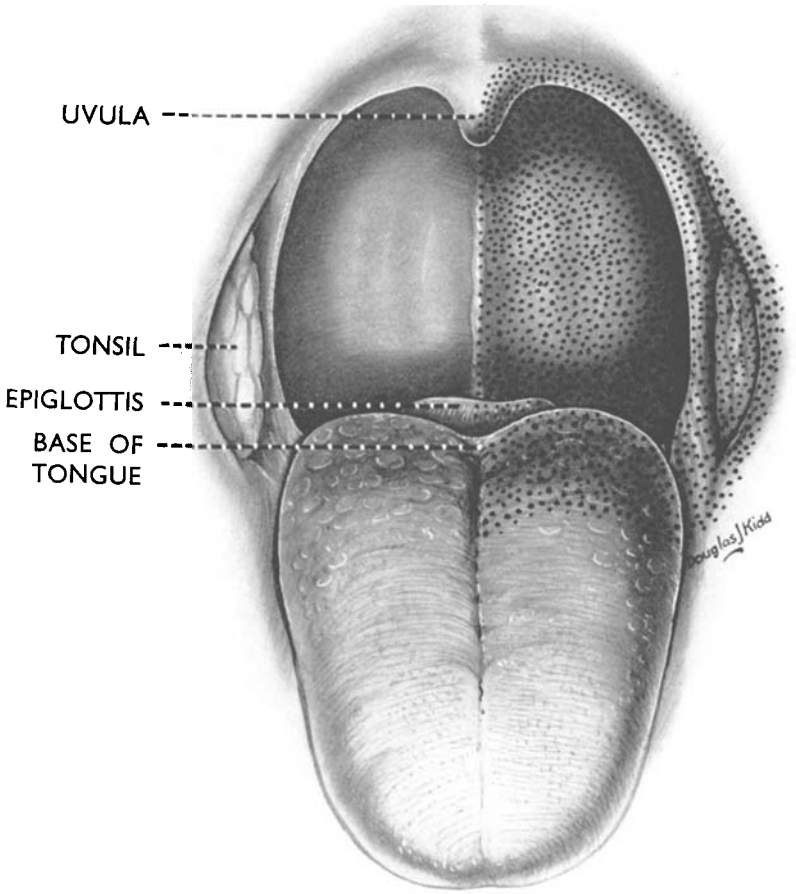


Fig. 2.

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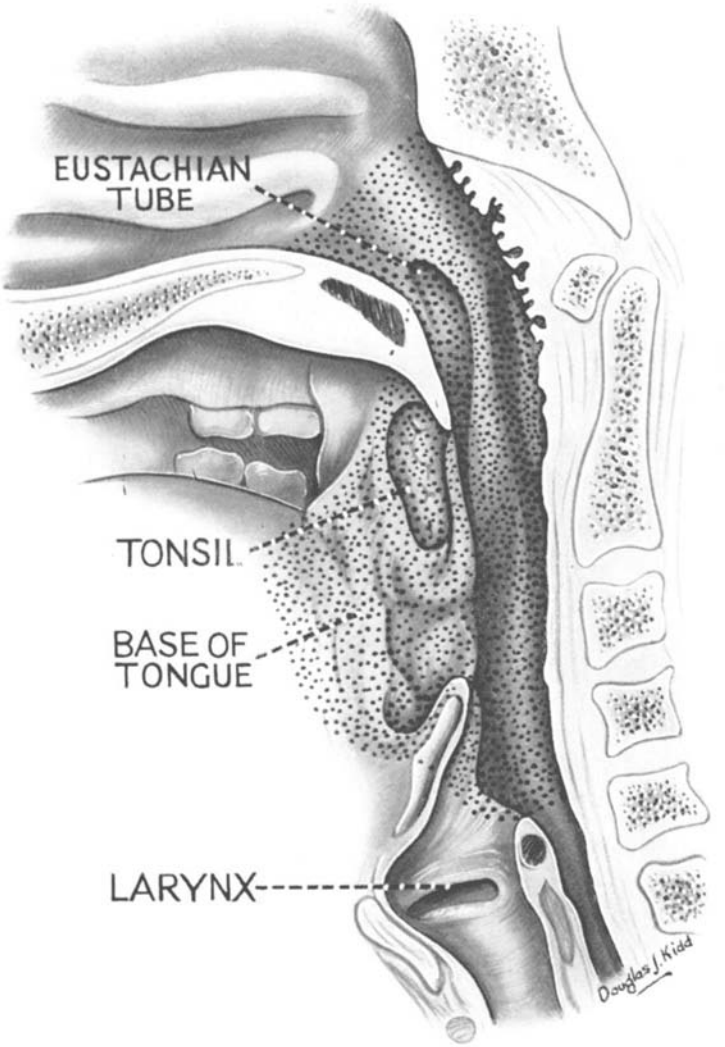


FIG. 3.

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been no recurrence of the pain but he now has slight analgesia over the right tonsillar area, posterior third of tongue and the right pharyngeal wall, including the orifice of the Eustachian tube; there is, however, no motor paralysis. The patient is now quite well and free from recurrence of the pain (June 2nd, 1937).

I would emphasize in this case that *extra-cranial* avulsion of the nerve gave over four years freedom from pain, but that *intra-cranial* section was necessary for a radical cure.

Case II.—Mrs. Y., P.P. B.39/15. Aged 57 years when first seen on November 28th, 1931. She gave a six years' history of paroxysmal acute stabbing pain in the left neck, tonsillar area and deep in the ear on swallowing or yawning. She said that her first swallow produced agonizing pain but that subsequently she was able to swallow without discomfort. She had seen her doctor in 1929 and told him that she felt sure there was a bone sticking in her throat, although no abnormality of the naso-pharynx was discovered nor was there any evidence of organic disease in the nervous system. An *extra-cranial* avulsion of the left glosso-pharyngeal nerve was performed in December, 1931, by Mr. Woolfenden, and since then she has had no recurrence of her pain (October, 1936).

Here then are two typical cases of primary glosso-pharyngeal neuralgia, the first demanding intra-cranial section for its radical cure, the second being still free from pain nearly five years after an extra-cranial avulsion. I have in all records of eight such cases, two of which have required intra-cranial section. In each, pain was complained of in the distribution of the glosso-pharyngeal nerve, i.e. the root of the tongue, tonsil, deep below the angle of the jaw, deep in the ear, etc.; and precipitating factors were found to be swallowing, especially of sour or sweet foods, chewing, talking, yawning, coughing, sneezing, stooping, speaking, syringing the ear, pulling the ear, examining the throat, etc. "Trigger zones" were found in the tonsil, the pharyngeal wall, the base of the tongue and the ear.

The *treatment* of primary glosso-pharyngeal neuralgia is essentially surgical. Medical measures are simply palliative. Sedatives and analgesics such as butyl-chloral hydrate, gelsemium, aspirin, codein, etc., can be given but rarely prevent severe paroxysms. Trichlorethylene which possesses the peculiar property of producing anæsthesia in the distribution

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of the Vth and IXth cranial nerves often relieves pain temporarily when 20-25 drops of a chemically pure preparation are poured on to gauze and inhaled through the nose two or three times a day; as transient faintness sometimes results from this, the patient should inhale the drug whilst reclining. The pain, however, can rarely be controlled by medicinal measures. Cure demands interruption of the sensory fibres of the glosso-pharyngeal nerve, proximal to the superior or jugular ganglion. Alcohol injection, which is so useful a procedure in trigeminal neuralgia, is impossible to carry out safely on the glosso-pharyngeal nerve because of its proximity to such important structures as the vagus, the jugular vein, etc.; moreover it is a small nerve with no adequately definable landmarks. Extra-cranial avulsion is a simple though somewhat hazardous procedure; that its effects may not be permanent, is illustrated by Case I, and if the petrosal ganglion is not avulsed, its tympanic branch may remain unsevered and thus pain associated with the ear and tympanum will persist. The method of choice is intra-cranial resection of the nerve; this offers the best chance of permanent cure. In expert hands it is not a formidable procedure; moreover, it excludes the remote possibility of an associated intra-cranial neoplasm such as an acoustic neuroma being overlooked.

An interesting phenomenon following section of the glosso-pharyngeal nerve is a transient rise of blood pressure. This results from cutting off the impulses ascending from the carotid sinus in the branch which joins the glosso-pharyngeal nerve below the ganglia; thus, depressor influences are partially removed and the blood pressure rises. This fact has an important bearing on the operative risk in cases of hypertension suffering from glosso-pharyngeal neuralgia, but as the effect is temporary, care will usually obviate the dangers of a cerebral vascular accident.

As an example of a secondary glosso-pharyngeal neuralgia, the following case is instructive.

CASE III.—Mr. J.B., R.I. 30057. Aged 63 years when admitted on December 21st, 1931. His history was that a year previously he had suffered from "tonsillitis" with difficulty in swallowing. This gradually disappeared but the left side of his throat remained sore and about the same time he experienced pain deep in the left ear, sharp and shooting in type, occurring at first especially on

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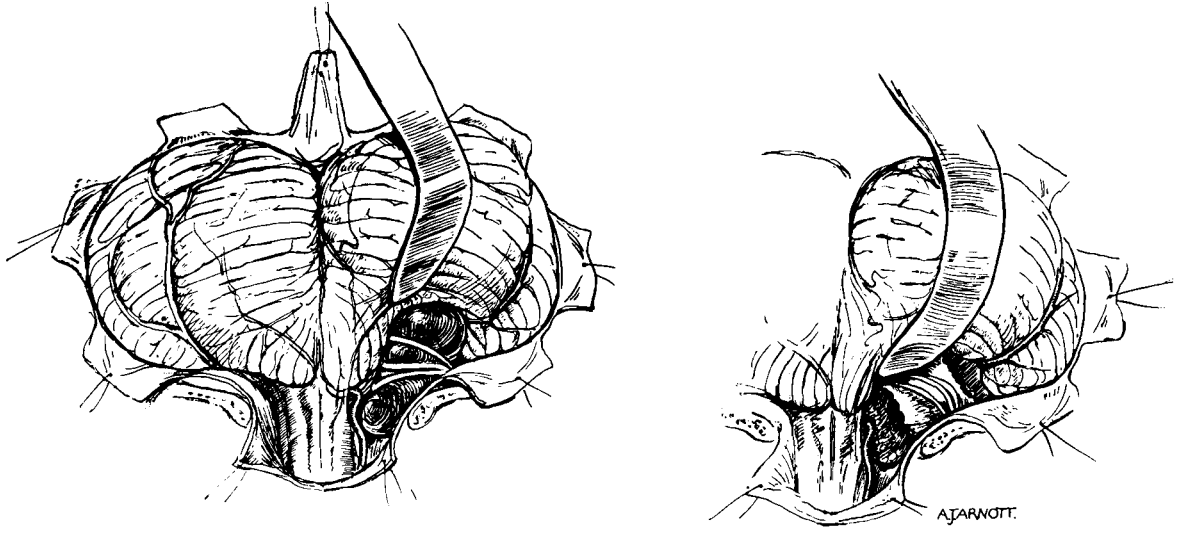


Fig. 4.

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swallowing but soon becoming more or less continuous. "I can occasionally swallow a mouthful of food in comfort but usually it touches a spot on the left side of my throat which feels as if something were tearing out the side of my throat and this starts the pain in my ear." Examination showed no abnormality except slight enlargement of the left tonsil.

On December 22nd, 1931, the left glosso-pharyngeal nerve was avulsed extra-cranially. There followed immediate and complete freedom from pain but this lasted two months only. Towards the end of February the pain recurred and was constantly present, gradually becoming more intense. The patient then noticed a small lump (an enlarged gland) below the upper part of the left sterno-mastoid. He was readmitted to hospital. Most careful re-examination failed to reveal a primary growth in the nasopharynx or upper respiratory tract, though an excised gland showed widespread invasion by an ill-differentiated squamous carcinoma.

In this case the character of the pain and the absence of complete remissions suggested a secondary neuralgia which was confirmed by the very temporary relief following avulsion of the glosso-pharyngeal nerve, and the subsequent appearance of secondary malignant glands.

Another interesting case of secondary glosso-pharyngeal neuralgia which I recall was associated with lymphadenoma and a Pel-Ebstein syndrome. In this case, a boy of 19 years, there was a history of recurring attacks of constant aching pain in the distribution of the right glosso-pharyngeal nerve lasting 8 to 12 days, and accompanied by fever and enlarged upper deep cervical glands. X-ray treatment to the site of the affected glands gave temporary relief. It was interesting to note the periodicity of the neuralgia, the exacerbations of which conformed to the febrile attacks, and the associated increase in the size of the cervical glands.

Secondary neuralgia results from gross pathology; but a gross lesion can involve a nerve without neuralgia necessarily resulting. The following case of *neurinoma of the glosso-pharyngeal nerve* was unassociated with a typical neuralgia.

CASE IV.—R.E., R.I. 43294. Aged 48 years on admission to L.R.I., January 15th, 1935. In April, 1934, he first experienced transient dizziness; in July, 1934, whilst in New York he had noticed a tendency to fall to the right on walking, with weakness and unsteadiness of the right leg. The following month whilst in Australia he felt pain down his left leg with stiffness and numbness

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of the right side of the face ; this gradually increased, lasted about three months, then gradually subsided ; he had occasional bursting headaches, and had noticed double vision, with increasing deafness in the right ear ; there was also weakness and unsteadiness of the right arm. He was investigated in Australia and sent back to England. For three months prior to admission he had complained of headache, maximally occipital, and had vomited on a few occasions. He had " curious sensations " on the right side of his throat and occasional " choking when swallowing fluids " ; articulation was sometimes difficult but this was a very variable complaint.

He was an intelligent co-operating patient, with a slow but distinct articulation. His fundi were clear, and his visual fields full but his visual acuity was diminished (left 6/24 ; right 6/18). The pupils were normal and ocular movements were full though there was well marked nystagmus in all directions, coarser on looking to the left. There was diplopia on looking upwards and to the right. The right corneal reflex was diminished and there was relative analgesia of the ophthalmic and maxillary division of the right Vth nerve. Taste was impaired in the anterior two-thirds of the tongue on both sides, especially the right. There was slight weakness of the right lower face and slight nerve deafness on the right side, with impaired caloric response from both labyrinths. The palate moved slightly to the right. The head was kept with chin to the left and raised. The right upper limb showed marked asynergia, hypotonia, ataxia and intention tremor ; these signs were present though less marked in the right lower limb. The gait was unsteady, with rather high stepping and a tendency to fall to the right. There was no gross sensory or reflex change in limbs or trunk and no sphincter disability.

His cerebrospinal fluid pressure measured (January 16th, 1935), 140 mm. ; the fluid was clear ; protein 180 mgm. per cent. ; cells, two lymphocytes; gold curve, 00111232100; Wassermann reaction, completely negative in all dilutions. The blood Wassermann reaction was negative. X-ray of the skull showed no gross change but there was slight thinning just behind the foramen magnum. Ventriculography (February 26th, 1935) showed symmetrical dilatation of the lateral ventricles and of the third ventricle ; the ventricular fluid showed no gross increase of protein. He had glycosuria due to a low renal threshold. His blood pressure was 120/85, pulse rate 72. There was no disease elsewhere.

The provisional diagnosis was a posterior fossa tumour involving the foramen magnum on the right side but the patient refused operation because he felt he was improving. He was seen by Mr. Hugh Cairns on March 15th, 1935, who confirmed the diagnosis but advised temporizing in view of the patient's improvement. A month later however he was no better ; the ataxia in his right

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arm had increased and he had less control of his right leg. On May 27th, 1935, Mr. Hugh Cairns operated and found a tumour in the right cerebello-pontine angle, low down and far forward (see Fig. 4). It was lying in front of the IXth and Xth nerves which were splayed across its middle. The tumour measured $3\frac{1}{2}$ cm. × 3 cm. × 3 cm., and appeared to be attached to the posterior part of the jugular foramen in the region of the IXth nerve whence it spread medially and forwards. It was fairly closely attached to the right restiform body. A fairly complete intra-capsular removal of the tumour was made, conserving the Xth but dividing a good portion of the IXth nerve. The patient stood the operation well.

The tumour contained two cysts full of bright yellow fluid. Examination proved it to be a *neurinoma*. When last seen (March 3rd, 1937) the patient was much improved.

Here then is a case in which a neurinoma involving the glosso-pharyngeal nerve was unaccompanied by a characteristic neuralgia.

In conclusion, I would reiterate the basic principles which are applicable not only to glosso-pharyngeal neuralgia but to all neuralgias: (1) there is a type of nerve pain unaccompanied by any demonstrable pathological change in the nerve or its central connections which has typical clinical features; this neuralgia should be called a *primary* neuralgia; (2) the pain which results from direct invasion of a sensory nerve by a gross lesion differs in type, evolution and accompanying signs from the primary neuralgia and should be termed a *secondary* neuralgia; and (3) the absence of pain does not exclude irritation of a sensory nerve.

If these principles are recognized and the terminology I have suggested is adopted, much of the confusion which at present clouds our understanding of the neuralgias should be dissipated.

. . . .

Primäre und sekundäre Neuralgien im Gebiete des Nervus glossopharyngeus sind viel schwerer zu erkennen, als diejenigen des Nervus trigeminus, weil objektive Störungen der Nervenfunktion nicht leicht nachweisbar sind. Dir Krankengeschichten verschiedener Fälle werden wiedergegeben. Dabei wird ausgeführt, dass in einem Falle trotz der extrakraniellen Evulsion des Nervs mit darauffolgender vierjähriger Schmerzfreiheit, zur völligen Heilung schliesslich doch ein intrakranieller Eingriff notwendig war.

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Professor Henry Cohen

Er glaubt, dass für alle Neuralgien folgende Grundsätze gelten :

(1) Es besteht eine Art von Nervenschmerz, der ohne nachweisbare Veränderungen des Nervs oder seiner zentralen Verbindungen doch ein typisches klinisches Bild darstellt. Dafür sollte die Bezeichnung " primäre Neuralgie " verwendet werden.

(2) Schmerzen, die durch direkte Erkrankung eines sensiblen Nervs durch gröbere Schädigung hervorgerufen sind, unterscheiden sich in der Art, der Entwicklung und den Begleiterscheinungen von der primären Neuralgie und sollten als " sekundäre Neuralgie " bezeichnet werden.

(3) Das Fehlen von Schmerzen schliesst die Reizung eines Sinnesnerven nicht aus.

Verfasser glaubt, dass bei Berücksichtigung des Gesagten die Verwirrung, welche in Bezug auf die Neuralgien besteht, verschwinden würde.

. . . .

Parce que les signes objectifs sont difficiles à démontrer, les névralgies primaires et secondaires de la lXème paire sont beaucoup plus difficiles à distinguer que celles dans la région du trijumeau. Les observations cliniques sont citées et on signale que quoique l'avulsion extracranienne donne un répit de douleur de plus de quatre ans, une cure radicale ne serait assurée que par une avulsion intra-cranienne.

L'auteur estime que les principes fondamentaux qui s'appliquent à toutes les névralgies sont (1) il y a un type de névralgie seule sans changement démontrable soit dans le nerf soit dans les noyeaux centraux : on appellerait ceci une névralgis primaire, (2) la douleur qui est le résultat d'une invasion directe d'un nerf sensoriel par une lésion, diffère en type, en évolution et en signes cliniques de la névralgie primaire : on appellerait ceci une névralgie secondaire, (3) l'absence de douleur n'exclut pas l'irritation d'un nerf sensoriel.

Il estime que si ce fait est accepté, beaucoup de malentendus au sujet des névralgies pourraient être vités.

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