Glossopharyngeal and Vagal Neuralgia

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Tic douloureux involving the glossopharyngeal nerve is rare and when it does occur probably often involves the vagus nerve as well. Many cases are undoubtedly not recognized, and yet the condition responds favourably and permanently to intracranial section of the glossopharyngeal nerve and upper filaments of the vagus nerve. We therefore propose to review the literature, discuss our own experiences, and try to clarify the clinical syndromes that this condition presents and the surgical approach that is required.

Review of the Literature

The condition of glossopharyngeal neuralgia was first described by Weisenburg (1910), who recognized it in a patient with a tumour of the cerebellopontine angle. In most cases, however, there is no known pathological condition. Harris (1926) described it as the idiopathic glossopharyngeal neuralgia. The usual description is of paroxysms of pain of a stabbing or lancinating character located in the base of the tongue and faucial region on one side, and provoked by swallowing, talking, and coughing. When the pain is severe the patient may not even be able to swallow his own spittle. Bouts of recurrent pain may last a few minutes, hours, or even days at a time, each individual stab, however, being momentary. The majority of patients are elderly, and in our series of 10 cases the ages ranged from 32 to 77 (average 62.6 years). Males are more often affected than females (9 to 1 in our series). Similar observations were made by Dandy (1927), but Bohm and Strang (1962) reported an equal occurrence in the sexes. As time goes by, however, recurrent bouts of pain tend to become more frequent and the periods of remission shorter.

One of the largest series so far reported is that of Bohm and Strang (1962), who made the point that the pain could radiate in other directions, notably into the vagal territory. This has been our experience also. Quite often the stabs of pain are felt deep in the external meatus of the ear and beneath the angle of the jaw, and sometimes even deeply in front of or behind the external auditory meatus. In some cases it may reach the tragus of the ear (with a trigger area there), but the main site of the pain is deep. When this occurs differential diagnosis from atypical forms of third-division tic douloureux becomes difficult (see below).

The surgical treatment reflects these difficulties in diagnosis as well as the efficacy of the various surgical approaches. There are three approaches, which may be described as cervical, tonsillar, and intracranial. The first successful approach was that of Sicard and Robineau (1920), who carried out a dissection of the glossopharyngeal nerve and pharyngeal branch of the vagus nerve in the neck and avulsed them. Harris (1926) was uncertain of the involvement of the vagus nerve and wondered whether section of the pharyngeal branch of the vagus nerve was necessary. Dandy (1927) also at first suggested that the vagus nerve is not implicated, and later advocated the intracranial approach with section of the upper filaments of the vagus nerve in addition to the glossopharyngeal nerve (Dandy, 1945). Adson, in 1925, was the first to use this approach, but his intervention was first reported by Love (1944). Dandy (1945) did record two cases in which, after intracranial section of the glossopharyngeal nerve alone, pain persisted, but it disappeared after a second operation in which the upper vagal rootlets were cut, an experience which Bohm and Strang (1962) also shared. Wilson and McAlpine (1946) reported a single case in which the glossopharyngeal nerve had been dissected in its tonsillar bed and avulsed with relief. Our experience, however, suggests that recurrence can arise after this procedure, and has led us to recommend intracranial section of the glossopharyngeal nerve plus section of the upper two vagal rootlets.

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Present Series

This study is based on a review of 10 patients admitted to this unit for treatment of glossopharyngeal neuralgia, with reference to type of pain, its aggravating factors, surgical treatment, and other relevant observations (see Table).

Duration and Type of Pain

The length of history varied from 15 months to 15 years. The pain was paroxysmal, and was described as lancinating or lightning-like jabs. The paroxysms occurred in characteristic bouts lasting from a few seconds to two minutes. Dandy (1927) wrote that the pain rarely lasted over a minute. In one of our cases, however, some bouts lasted up to five minutes, and paroxysms of pain continued to occur at frequent intervals throughout the day over a period of a few days to a few weeks before a spontaneous period of remission ensued. The pain is very severe, and in Case 3 the paroxysms occurred so often that section of the glossopharyngeal nerve was performed as an emergency to enable the patient to swallow. The period of remission before operation was also variable in this series: the longest was four years (Cases 1 and 4). Adson (1924) reported a remission of three years and nine months. One observation which stands out in this series is that the periods of remission become shorter with ensuing attacks, but an identical pain always returns in the same area of distribution.

The location and distribution of pain in this series was always unilateral, and was as follows (see Table): back of tongue (Cases 9 and 10), fauces (Cases 3 and 10), tonsillar region (Case 7), deep in the ear (Cases 1, 2, 5, and 6), in front of or behind the ear (Cases 2 and 5), deep to the angle of the jaw (Cases 3, 5, 6, and 8), deep in the upper side of the neck (Cases 1, 4, and 8), and deep in the throat (Cases 1 and 2). Only one patient (Case 9) had pain confined to the back of the tongue with no radiation.

Aggravating Factors

The commonest method of precipitating attacks of the pain was swallowing, but attacks were also induced in other ways, such as coughing, sneezing, talking, touching the tragus of the ear, turning the head towards the side of pain, and rolling towards the site of pain.

In only one patient (Case 4) was there a trigger zone. His pain was brought about by touching the tragus on the painful side, but in three other patients the tragus was also sensitive.

Other Observations

There was no seasonal influence on the symptomatology, except in one patient (Case 1) who had pain only in winter. One patient (Case 2) had vertigo without tinnitus or deafness. Bohm and Strang (1962) reported two similar cases. None of our patients had any sensory impairment in the area of the distribution of pain. All were symptom-free in between attacks of pain, apart from one patient (Case 1) who complained of a hot flushed feeling in the affected area between the bouts of pain. Our follow-up periods (excluding one patient (Case 7) who died postoperatively) have ranged from four and a half weeks to six years, and all patients appear to have been relieved during that time.

Treatment

Seven out of the ten patients were treated by section of the glossopharyngeal nerve and the upper rootlets of the vagus nerve, and two by section of the glossopharyngeal nerve alone. One patient (Case 10) was pain-free on admission, but was admitted later to another neurosurgical unit with recurrent pain. He was operated on and remained free from pain until he committeed suicide because of depression. The postoperative death, on the 11th day, on a 77-year-old patient, was due to intestinal obstruction (Case 7). Seven out of the remaining eight patients with an adequate follow-up have remained pain-free. The eighth patient (Case 9) originally had pain in the back of the tongue on one side, and after operation was free of pain for four months. Pain then occurred in the lower jaw. He was treated by alcohol injection of the trigeminal ganglion and has since remained pain-free. It is possible that he had both glossopharyngeal and trigeminal neuralgia. Bohm and Strang (1962) described a similar case in which second division trigeminal neuralgia occurred five years after a glossopharyngeal section.

Postoperative Course

Only one patient (Case 4) had transient difficulty in swallowing. Six patients showed detectable sensory depression in the oropharynx, the faucial region, the posterior third of the tongue, and the adjacent half of the soft palate. Two patients had sensory depression in the concha, one of them having had no sensory depression in the oropharynx. In one case no mention of any sensory loss was recorded. Another patient (Case 3) developed transient auricular flutter after he had undergone glossopharyngeal section with the upper rootlets of the vagus.

Discussion

Compared with trigeminal neuralgia, glossopharyngeal neuralgia is relatively rare. Bohm and Strang (1962) showed an incidence of 1% in Olivecrona's material, and Harris (1937) an incidence of 0.57%. The corresponding figure for this neurosurgical unit is 0.75%.

The presence of a trigger zone is a rare finding, though there may be areas tender and sensitive to touch. One case in our series involved the tragus (Case 4). Harris (1926) remarked on the absence of trigger zones and thought it to be the main difference between glossopharyngeal and trigeminal neuralgia. However, cases with a trigger zone in the faucial region have been described (Dandy, 1927).

The location, distribution, and radiation of pain lie in the area of sensory supply of the glossopharyngeal nerve and of the auricular and pharyngeal branches of the vagus nerve—i.e., propharynx, concha of the ear, and deep to the angle of the jaw. Dandy (1927) implicated Jacobson's nerve in the radiation of pain to the ear, and described the radiation of pain to the angle of the jaw and concha as referred. Keith (1932) described pain in the regions of the upper part of the neck, the angle of the jaw, and the cheek, nose, and eyes, and regarded them as overflow phenomena. We think that pain deep to the angle of

Salient Features of Cases in Chronological Order

Case No.	Age and Sex	Duration of History of Pain Before Operation	Chief Site of Pain	Radiation	Aggravating Factor	Trigger Area	Treatment (Intracranial)	Duration of Follow-up	Results and Remarks
1	59 F	9 years	Deep in ear	Deep in side of neck	Swallowing. Washing concha		Division of 9th nerve and upper two rootlets of vagus	6 years	Section of 9th cranial nerve in neck 9 months previously. Pain-free since intracranial operation. Sensory depression faucial region and posterior part of tongue
2	50 M	7	Deep in ear	Throat and behind ear	Swallowing. Coughing			5	Pain-free. Sensory depression in concha
3	76 M	3	Faucial region	Angle of jaw	Swallowing. Talking			2 ,,	Death due to acute abdomen; pain-free for the period
4	57 M	10	Deep in side of neck at end of thyroid cartilage	Ear	Swallowing	Tragus		3 ,,	Pain-free. Sensory depression in concha
5	64 M	15 months	Deep in ear	Back of ear and angle of jaw	Swallowing	Tragus sensitive to touch		4	Pain-free. Sensory depression faucial region and soft palate
6	77 M	5 years	Deep in side of throat	Deep in angle of mandible and deep in ear	Swallowing. Coughing. Turning head to side of pain	Tragus sensitive		2	Pain-free. Sensory depression faucial region and posterior part of tongue
7	77 M	15	Tonsillar region	Nil	Swallowing. Coughing. Eating		Division of 9th nerve		Had section of 9th nerve in tonsillar bed 14 years before admission and remained pain-free for 12 years. Death on 11th postoperative day from bronchopneumonia and intestinal obstruction
8	32 M	2	Tragus	Deep to angle of mandible other side of throat	Swallowing. Talking. Brushing teeth	Tragus sensitive	Section of 9th nerve and upper two rootlets of vagus	4½ weeks	Did not turn up for follow-up. Pain-free postoperatively, but address unknown
9	61 M	2	Back of tongue		Rolling of tongue to side		Section of 9th nerve	18 months	Pain recurred in 3rd division of trigeminal; has remained pain-free after alcohol injection
10	73 M	4½	Faucial region	Back of tongue			Section of 9th nerve at another neurosurgical unit	3 ,,	Pain-free till suicide because of depression

the jaw, upper cervical region, and concha is due to involvement of the auricular branch of the vagus nerve. We did not observe overflow to the cheek, nose, or eyes. Bohm and Strang (1962) subdivided their series into two groups—oropharyngeal and aural—according to the main symptomatology, but patients usually present with pain in both zones concurrently. Typically there is no sensory impairment in the area of pain, as there usually is when tumours of the cerebellopontine angle or base of the skull are present.

The clinical picture of "idiopathic glossopharyngeal neuralgia" is so characteristic that diagnosis is seldom difficult. Glossopharyngeal neuralgia, however, may be confused with trigeminal neuralgia in patients with pain in the region of the tragus or deep to the angle of the jaw.

There is some evidence that glossopharyngeal neuralgia may be controlled by medication—for example, vitamin B complex, Tegretol (carbamazepine)—but no proof that the relief is lasting. Treatment of this neuralgia is essentially surgical. It has passed through various stages from dissection and avulsion of the glossopharyngeal nerve and pharyngeal branch of the vagus nerve in the neck to the intracranial section of the glossopharyngeal nerve alone or combined with the upper two rootlets of the vagus nerve. However, the avulsion of nerves in the neck has often been followed by recurrence of pain over a period of time—for example, Case 1—as has avulsion in the tonsillar fossa (Case 7). The treatment of choice remains the intracranial section of the glossopharyngeal nerve and upper two rootlets of the vagus nerve, as we feel that the latter nerve is also implicated. In skilled hands this operation carries a negligible mortality and can be performed on elderly patients.

Surgical Approach

This is essentially the approach by Dandy (1927), though nowadays it is facilitated by operating on the patient in the upright position. The legs should also be raised or bandaged in elderly patients to prevent postural arterial hypotension. The procedure is carried out through a small suboccipital craniectomy under endotracheal anaesthesia. Enough bone is removed to expose the lateral sinus above and the sigmoid sinus laterally—that is, about 3–4 cm. diameter. The dura is opened, with a small flap swung laterally over the sigmoid sinus. The cerebellopontine angle is opened up after the posterior fossa has been slackened either by slipping a retractor over the cerebellar hemisphere and opening the cisterna magna or by releasing a previously placed lumbar puncture needle. The rootlets of the ninth, tenth, and eleventh cranial nerves are seen arranged in a vertical column from above down. The ninth cranial nerve is a single filament normally separated from the multiple filaments of the tenth nerve by a dural isthmus as the nerves pass out through the jugular foramen. The ninth cranial nerve is hooked up and sectioned at this position, followed by section of the upper two rootlets of the vagus. In closing the craniectomy the dura is sutured back into position. The patient is usually sitting out of bed on the first day, and relieved of pain from the time of operation.

Summary

Ten cases of glossopharyngeal neuralgia have been studied and the literature has been reviewed. A more correct classification would be to describe the condition as "glossopharyngeal and vagal neuralgia." Evidence is given to support this view. Intracranial section of the glossopharyngeal nerve and the upper two rootlets of the vagus nerve is the operation of choice.

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Haemoglobin F Hull (γ 121 Glutamic Acid → Lysine), Homologous with Haemoglobins O Arab and O Indonesia

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A haemoglobin variant is the result of an amino-acid substitution in one of the two pairs of polypeptide chains which constitute the haemoglobin molecule. Foetal haemoglobin (Haemoglobin F) has a pair of α-chains and a pair of γ-chains, and is denoted as $\alpha_2\gamma_2$. Variants of Haemoglobin F may thus be divided into two groups, those with abnormal α-chains and those with abnormal γ-chains.

An α-chain substitution gives rise not only to a variant of Haemoglobin F but also to a variant of Haemoglobin A ($\alpha_2\beta_2$) and a variant of Haemoglobin A_2 ($\alpha_2\delta_2$) in the same individual. A γ-chain substitution, in contrast, can give rise only to a variant of Haemoglobin F. In the same way a β-chain substitution can give rise only to a variant of Haemoglobin A ; most of the common haemoglobin variants—for example, Haemoglobins S, C, D, and E—are β-chain variants.

Few γ-chain variants have been described, and in only one case, that of Haemoglobin F Texas I, has the amino-acid substitution been identified (Schneider and Jones, 1965 ; Jenkins, Beale, Black, Huntsman, and Lehmann, 1967). We here describe a new γ-chain variant and define its amino-acid substitution. The variant, which is designated Haemoglobin F Hull, was first found as an electrophoretically slow component of the cord-blood haemoglobin of a normal baby of native English stock (Fig. 1). It made up 14% of the total haemoglobin, determined by the method of Marengo-Rowe (1965). It was found again in the cord blood of a subsequent baby

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