GENICULATE NEURALGIA

REPORT OF A CASE RELIEVED BY INTRACRANIAL SECTION OF THE NERVE OF WRISBERG*

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(Received for publication February 18, 1950)

Our teacher and friend, whom we are gathered here today to honor, has long been distinguished for his interest in the studies of the motor division of the 7th cranial nerve. It seems, therefore, particularly appropriate that your speaker, as one of his former students, should present a case involving the sensory division of this nerve.

It is my belief that the patient in this case has been correctly diagnosed as having geniculate neuralgia and that she has been relieved of her pain through intracranial section of the pars intermedia of Wrisberg. According to medical writing, this is one of the rarest of neurosurgical procedures. Because of the complexity of the sensory supply to the region of the ear and the rarity of the syndrome here presented, it might be of interest to review briefly some of the high lights in the evolution and development of our present knowledge of the functions of the 7th nerve.

Its anatomy as we know it today was described by Eustacius early in the 17th century. Sir Charles Bell was the first to bring this nerve into clinical prominence by his studies on facial palsy.

For a long time the 7th nerve was regarded as purely motor in function. From time to time an occasional sensory symptom has attracted attention, but such sensory manifestations have been referred to as produced by one of the many neighboring systems that converge and anastomose in this region.

In 1876 Webber carefully described the pain phenomena observed in 6 cases of facial palsy. The pain was located in the ear and mastoid regions and in some cases, radiated to the face and occiput. He thought the pain was mediated through the trigeminal and the auricular branch of the vagus. Cushing observed a crude sort of sensation persisting in the anterior two-thirds of the tongue, after posterior root section of the gasserian ganglion, in spite of anesthesia to touch, pain and temperature; later in one of his cases a facial palsy supervened and this crude sensation then disappeared, indicating general sensory fibres in the chorda of facial origin.

Spiller observed that light touch, pain and temperature sensations were lost in the trigeminal area after gasserian ganglion extirpation, but that deep pressure sensations were retained.

* Presented May 30, 1949 at a meeting of his former Residents, honoring Doctor Claude C. Coleman and commemorating the Thirtieth Anniversary of the establishment of the Department of Neurological Surgery, Medical College of Virginia, Richmond.
The investigations of His, Retzius, Sapolini and others have conclusively shown that the facial nerve has a sensory ganglion, the geniculate; a sensory root, the pars intermedia of Wrisberg; as well as sensory fibres coursing in the chorda tympani, the great superficial petrosal nerve and the trunk proper of the 7th.

Ramsey Hunt made his first report in 1907 on herpetic inflammations of the geniculate ganglion and for the next 30 years elaborated on the subject through numerous contributions, which are responsible for our present knowledge of this highly complex system. Much of this knowledge was acquired through his study of herpes of the geniculate ganglion and he was thus able to map out the cutaneous zone of innervation of the geniculate ganglion into what is known as the “zoster zone,” which includes the concha, tragus, antitragus, lobule and anthelix, the external auditory canal and a portion of the tympanum.

The pain in this condition is localized chiefly in the depths of the ear and in certain areas of the external ear, within the zoster zone. The pain is often lancinating and very severe and extends into the mastoid and occipital regions and into the face. These observations led to the recognition of the geniculate ganglion syndrome and established the fact that the facial nerve has a definite sensory system. Like the other mixed cranial nerves, the geniculate ganglion is composed of unipolar cells, the central processes of which terminate in the fasciculus solitarius of the medulla, with the central processes of the 9th and 10th, and constitute its sensory root. Hunt then summarizes his views on the geniculate system as follows:

1. Sensory filaments to the internal ear and branches to the zoster zone of the auricle, which bring it into close relation with the auditory mechanism. These branches are responsible for the otaiga.

2. The sensory system of the great superficial petrosal nerve, which brings the geniculate ganglion into close relation with the orbital, nasal and palatal branches of the sphenopalatine ganglion and the maxillary division of the fifth nerve. These branches are responsible for the deep prosopalgia.

3. A viscerosensory system subserving deep sensibility of the face, which is the seat of painful pressure sensations in geniculate prosopalgia.

These various branches of the sensory facial system have numerous anastomotic connections with the trigeminal, glossopharyngeal and vagal systems, as well as with the branches of the cervical plexus.

Not only are there these superficial connections, but the central sensory nucleus of the facial nerve stands in close anatomic relationship with those of the other mixed cranial nerves.

The central and peripheral associations with neighboring sensory systems account for the wide diffusion of pain in severe geniculate neuralgia.

It should be added that the facial nerve is especially rich in sympathetic and parasympathetic fibers, which explains the associated vasomotor and secretory manifestations.

Neuralgia facialis vera is characterized by pain in the distribution of the various divisions of the geniculate system. It may manifest itself as earache and as deep facial neuralgia. Geniculate prosopalgia is characterized by deep-seated pain in
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the face in the posterior orbital, nasal, malar and palatal regions, associated with pressure pain sensations. There are associated otalgia and referred pains in the mastoid and the occipital region, at times extending to the neck and the shoulder.

In some cases geniculate otalgia predominates; in others deep prosopalgia is more in evidence. This is in harmony with neuralgia of the other mixed cranial nerves—the fifth, ninth and tenth—in which partial and incomplete forms are encountered.

I have used the term deep prosopalgia not only because it expresses the essential nature and localization of the pain but in order to place it in contrast with the prosopalgia of trigeminal origin, which is more superficial and with which it has been confused.

CASE REPORT

This patient had 5 hospital admissions under my care and 1 at a hospital in Detroit under neurosurgical care.

1st Admission. She was first admitted to the Charleston General Hospital, July 1, 1943, at the age of 46, having been referred by Doctor Burian of McVeigh, Kentucky.

She complained of pain in her right ear and cheek which had come on suddenly 5 weeks previously while she was waxing her floors. The pain appeared to be deep in the ear and radiated to the eye, lips and nose, and lower jaw. It had been constant and continuous since its onset and was not aggravated by talking, washing face, eating or swallowing.

Previous operations were appendectomy, cholecystectomy, sterilization and bilateral mastectomy for benign lesions of the breasts. She had had “sick headaches” for years, benefited by Gynergen.

She was a large woman who appeared to be in excellent physical health. B.P. 160/100 (on subsequent visits it was perfectly normal). There were no objective findings of significance. X-rays of skull, and blood, urine, and CSF studies were negative. She was discharged without diagnosis; it was felt that she did not have major trigeminal neuralgia.

2nd Admission, October 1943 (3 months later). She had the same complaints of pain deep in ear, radiating to face. Again routine studies revealed no abnormal findings and she was discharged, undiagnosed.

3rd Admission, March 1946 (almost 3 years after the 1st). Her complaints were the same and the pain had continued without interruption since its onset 3 years previously. She thought that at times it was made worse by washing hair and face, so the 2nd division of the right 5th nerve was avulsed. She stated that this procedure relieved her pain and she was discharged.

4th Admission (9 months later). She stated that she had remained completely relieved until 2 weeks before readmission, when she was awakened from a sound sleep by a sudden, sharp pain deep in her right ear. It seemed to spread out into her right face and eye, and over the nose and downward over the right neck. There had been no let-up in the pain, which she described as dull and aching in the ear “as though it is going to burst.” She stated that the pain had been so “fierce” that she hadn’t been able to retain much food. She felt that there was a trigger zone in front of the right ear.

Since she had gotten complete relief for 9 months through avulsion of the right 2nd division, it was felt that we were dealing with a major trigeminal neuralgia after all, and a selective retrogasserian neurectomy was carried out in October 1946. This
gave complete and immediate relief and on her 3rd postoperative day she ate a good lunch with the comment that "this is my first good meal in 4 weeks," and a few days later stated that she felt "grand."

She remained free of all pain and nothing further was heard from her till 13 months later.

4th Admission (Detroit). A telegram was received from her daughter in Detroit requesting information for the hospital there. The patient had been suddenly seized with a violent deep right ear pain again and was hospitalized 10 days as a brain tumor suspect—but electro- and pneumoencephalography and other studies failed to reveal any evidence of a neoplasm. The opinion was expressed that she was suffering from a tension state with exaggeration of her symptoms.

She returned to her home in Kentucky and endured her pain with fortitude apparently for about a year, when there was an increase in intensity and a spread of her pain to the occipital region for the first time. During the next several weeks her husband spent, he stated, $50.00 for codeine, which she would use when her pain became unbearable. Many nights she was unable to sleep at all and would walk the floor crying out with her pain. It was in such a state that she reentered the hospital.

5th Admission, Mar. 13, 1949 (nearly 6 years after onset of pain). She had had no relief save for the 9 months following avulsion of 2nd division and 13 months following posterior root section of the 5th nerve.

She was quite obviously in very great anguish at this time. In addition to her deep ear and face pain she complained quite bitterly of tenderness in the occipital muscles, and the greater and lesser occipital nerves were infiltrated with a procaine ammonium sulfate solution without much relief. X-rays of cervical spine showed hyper tropic changes throughout.

After a week of observation and hours spent in talking with and examining her, the writer was firmly convinced of the genuineness of her complaints and of the necessity and desirability for a new approach to the problem, if relief was to be obtained. The clue was furnished by perusal of Ramsey Hunt's scholarly articles on geniculate neuralgia. Her symptoms appeared to be entirely in keeping with those of this malady.

On Mar. 22, 1949, operation was performed which she entered into with enthusiasm since it promised relief from her symptoms. Under local anesthesia a right unilateral subcerebellar craniotomy was done and the 7th, 8th, 9th and 10th cranial nerves were exposed. The 9th was touched first but because of the pre-operative Nembutal, her answers were not sufficiently clear-cut as to be dependable, so this nerve was severed. She stated that her old pain was still present and unchanged. The 8th nerve was then gently elevated and displaced backward with the shank of an angled hook knife, and the pars intermedia of Wrisberg picked up and sectioned. A definite answer that her pain had ceased was obtained. No bleeding was encountered and the dura and wound were closed.

A peripheral facial weakness developed postoperatively but since she still had corneal sensation, lid suture was not considered necessary. Otherwise her convalescence was uneventful. She left the hospital on her 9th postoperative day, entirely relieved of her former ear, face and neck pain. She reported for examination 6 weeks later and stated that she has had no suggestion of her former pain and her facial paralysis was clearing. She notes no especial change in her ear save that it is not quite as sensitive as the left though she does not recognize any numbness as such in the ear. Hearing was not impaired. She stated that during much of the time of her pain,
there was an excessive salivation so that she had considerable expectoration, nasal secretion and tearing and that all these symptoms have cleared since operation, save the tearing which was still present, presumably because of the facial paralysis.

A review of the literature reveals only 2 such patients operated on previously. The pain in both of the reported cases was paroxysmal rather than continuous.

The first case was reported by Clark and Taylor in 1909 and the diagnosis was made possible by Hunt’s earlier work. Their patient was a housewife, aged 28, who had had paroxysmal intermittent pain in front of the left ear for 2 years. About 8 months after onset, the pain had spread to involve the ear canal as well as the three branches of the 5th nerve and the occipital region. She was taking 12 gr. of morphine daily. Taylor severed the pars intermedia of the 7th nerve and the upper fasciculus of the 8th nerve with complete, immediate and permanent relief of pain with no further need or desire for opiates. This patient had a facial paralysis which was clearing 5 weeks later.

The second case report was that of Furlow who operated on a young woman in November 1941 because of a shooting pain in the left ear of some 2 years’ duration. Her pain was not continuous but would come and go spasmodically and was located chiefly deep in the ear, sometimes radiating to the face and neck. On one occasion cocainization of the sphenopalatine ganglion gave temporary relief. Apparently, this procedure interrupted a pain reflex as did the avulsion of the 2nd division and later, posterior root section, in our own case.

Reichert in 1933 reported the case of a telephone operator, aged 31, with a similar history of a severe, intermittent, paroxysmal pain in the left ear. At times she was unable to use ear-phones because this disturbed the trigger zone on the concha and set off her pain. At times she also had aching pains in the left side of nose, face, eyeball, parieto-occipital and mastoid areas. He thought this was a case also of geniculate neuralgia, but upon operation under local anesthesia, her identical pain was reproduced when the 9th nerve was touched, so it was sectioned with complete and immediate relief. He had first gently touched the bundle of the 7th and 8th nerves and the patient complained of ear pain, but it did not have the characteristics of her old pain. This, then, was a case of tic douloureux of the branch of the glosopharyngeal (Jacobson’s tympanic plexus branch). It is a variant of the glosopharyngeal neuralgia as it is typically found and illustrates again the complexity and overlapping in the mixed forms of otalgia in which the 5th, 7th, 9th and 10th cranial and the occipital nerves may play a part.

It is not surprising to have a partial neuralgia of a mixed cranial nerve. It is rather common to have a single division of the 5th nerve involved, later to spread to other divisions.

Since Reichert had thought he was dealing with a geniculate neuralgia and no doubt would have severed the nerve of Wrisberg had the patient been under general anesthesia, this illustrates the necessity of a local anesthetic
in such cases, as was first used by Davenport in a case of glossopharyngeal neuralgia. It should be invaluable for the confirmation of clinical evidence in a case in which the diagnosis is in doubt. Had the pre-operative Nembutal not robbed our patient of her ability to cooperate accurately, her glossophranygeal nerve probably would not have been sacrificed.

It is perhaps not surprising that the function of the sensory mechanism of the 7th nerve should so long have escaped detection. This is a small ganglionic system intercalated between the large system of the trigeminus in front and the systems of the glossopharyngeal and vagus and the upper cervical nerves behind. As a result, there are numerous anastomoses of which it is very difficult to determine their motor or sensory nature and their proper ganglionic origin. The problem is further complicated by the intricate relations of the facial system to the highly specialized structures of the auditory mechanism. That the distribution on the auricle was not recognized is also easily understood. The area is small and because of its situation in the auricular folds and the canal it is difficult of access. And further, the concentration of the auricular branches of the 5th, 9th, 10th and upper cervical nerves in the same region still more complicates matters, so that a small area of anesthesia in this region would rapidly fade in the overlapping of adjacent marginal areas.

In the matter of differential diagnosis, tic douloureux of the 5th and 9th nerves should cause no great difficulty as the symptomatology and treatment of these maladies is well understood and standardized. When one considers the great frequency of trigeminal neuralgia and the comparative frequency of glossopharyngeal and even of superior laryngeal (vagal) neuralgia, one should not expect an important sensory system like that of the geniculate to escape and no doubt many cases of this sort have masqueraded under the title of the atypical facial neuralgias.

A few years ago, a very distinguished visiting neurosurgeon, upon being asked to discuss atypical facial neuralgia, arose and stated simply: "I let them severely alone." He probably was being facetious but the problem isn't funny, either to the patient who has the malady or to the physician who assumes to treat such complaints and it behooves us all to take more seriously complaints of pain and make an effort to correctly diagnose them. Glaser, analyzing 143 cases of atypical neuralgia, made a careful analysis of the symptomatology. He concluded that the pain was not superficial. It is not referred to the surface as in trigeminal neuralgia but is deep seated in the tissues, the bone, and the eyeball. The pain does not follow the distribution of the various branches of the 5th nerve but jumps anatomic boundaries and extends into the neck and arm.

In addition to pain, about half of the patients studied had associated sympathetic phenomena, which suggested the sympathetic system as the origin of the pain in atypical neuralgia. However, the late and to us here the much lamented Dr. Max Peet concluded that atypical neuralgia could be due to sympathetic origin only through vasomotor spasm. Fay spoke of a
syndrome of vascular pain to account for atypical neuralgia, expressing the opinion that pain follows the course of the vascular tree. In one case he performed 7 operations in an effort to cure a patient of atypical neuralgia: (1) Section of the trigeminal root; (2) resection of the sphenopalatine ganglion and the upper branches of the 7th nerve; (3) ablation of the cervical portion of the sympathetic trunk below the superior cervical ganglion; (4) stripping of the common carotid artery; (5) section of the hypoglossal nerve; (6) extracranial section of the glossopharyngeal nerve; (7) section of the sensory branches of the vagus nerve, the jugular branch of the vagus and Arnold’s nerve and division of the sheath of the vagus and of vagus fibres to the internal and external carotid artery.

In Sluder’s syndrome, Sluder\textsuperscript{22} states “the pain of sphenopalatine neuralgia has a wide distribution and is localized in the root of the nose, in and about the eye and in the upper part of the face and the upper teeth and sometimes also in the lower jaw, the lower teeth, in the temple and about the zygoma, the ear, the mastoid, the occiput and the neck. It may extend to the shoulder and less often to the axilla and the breast, and in severe attacks, to the arm, the forearm, the hand and even the finger-tips.” He also described vasomotor and secretory symptoms referable to the sympathetic system such as sneezing, congestion of mucous membranes with hydorrhea, symptoms our patient had also.

Vail\textsuperscript{24} stated that vidian neuralgia is characterized by pain in the nose, face, eye, ear and sometimes neck and shoulder.

Ruskin\textsuperscript{29} concluded that the sphenopalatine ganglion syndrome of Sluder is “not a distinct clinical entity but is composed of four distinct syndromes: maxillary, sensory facial, sympathetic and local sphenopalatine (cells of the ganglion itself)”.

Such symptoms as the above should certainly be carefully scanned as possible cases of geniculate neuralgia.

**SUMMARY**

A case is presented in which the symptoms appear to be entirely in keeping with what Ramsey Hunt has described as geniculate neuralgia, mediated through the sensory portion of the 7th cranial nerve. Section of the pars intermedia of Wrisberg has given complete and immediate relief of a severe and continuous pain deep in the ear, face, neck and occipital region of 6 years’ duration, save for two periods of 9 and 13 months following avulsion of the 2nd division of the 5th nerve and posterior root section respectively. It is admitted that this statement is open to the criticism that not enough time has elapsed since operation to be sure of its permanency.*

And in closing, I would like to summarize the present status of neuralgic affections of the various cranial nerves in the words of Hunt.

1. True trigeminal neuralgia, which is distributed in one or more branches of the trifacial nerve and in which the pain is localized in the more superficial structures of

* Her complete relief continues in September 1930.
the face and intra-oral region. This is classic prosopalgia or trifacial neuralgia. In cases of neuralgia of the third division of the fifth nerve there is often associated otalgia.

2. Geniculate neuralgia, which involves the deeper structures of the face. This is characterized by pain in the deep posterior orbital, palatal and nasal regions, with painful pressure sensation in the face. This is geniculate deep prosopalgia, and with it there is associated geniculate otalgia.

3. Glossopharyngeal neuralgia, which is characterized by neuralgic pains in the distribution of the glossopharyngeal nerve at the base of the tongue and the adjacent regions of the throat and by associated otalgia.

4. Superior laryngeal neuralgia, of vagal origin, in which the pains are localized in the region of the larynx, with associated otalgia.

All these various forms are accessible to surgical intervention by the cranial method of approach through the posterior fossa, which exposes the fifth, seventh, ninth and tenth cranial nerves. If this procedure is carried out with the use of local anesthesia, it is possible by touching any one of the nerves to reproduce the neuralgic pain, thus confirming the clinical diagnosis, which is often involved and difficult.

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