TUMORS OF THE GASSERIAN GANGLION

TUMOR OF THE LEFT GASSERIAN GANGLION ASSOCIATED WITH ENLARGEMENT OF THE MANDIBULAR NERVE

A REVIEW OF THE LITERATURE AND CASE REPORT

HENRY M. CUNEO, M.D., AND CARL W. RAND, M.D.

Department of Neurosurgery, University of Southern California School of Medicine,
Los Angeles, California

(Received for publication March 7, 1952)

The trigeminal nerve and its ganglion may be the site of a variety of pathologic lesions. Primary tumors may arise either in the Gasserian ganglion itself or in the root of the trigeminal nerve. Meningiomas, neurofibromas, gliomas, ganglioneuromas, adamantinomas, neurocytomas, and metastatic carcinomas are some of the various tumors that have been described. In addition, sarcomas and osteochondromas of the mandible, antrum, nasopharynx or internal ear, have been reported. Aneurysms of the internal carotid artery may also cause symptoms similar to those of tumors. The clinical findings and symptoms that these lesions may produce may make localization of the lesion, whether in the ganglion itself or in the nerve root, relatively simple. On the other hand, it may be impossible at times to determine preoperatively whether the tumor is in the middle or the posterior fossa of the skull, or both.

Sachs,13 in 1917, writing of tumors of the Gasserian ganglion, mentioned that the earliest case was reported by Dixon in 1846, and one was reported by Günsburg in 1849. Peet,12 in 1927, found a total of 63 cases in the literature, and referred to a case studied as early as 1836 by R. W. Smith and reported in 1849. Peet described 2 cases of his own in which the tumors had infiltrated the Gasserian ganglion by direct extension through the maxillary division. In his first case the diagnosis was medullary squamous cell carcinoma, without cornification. In the second case, the maxillary division had been infiltrated extracranially by a squamous cell carcinoma of the right antrum. Microscopic examination of the sensory root and posterior portion of the ganglion showed no pathologic change.

Learmonth and Kernohan,10 in 1930, described a tumor of the Gasserian ganglion said to be similar to neurocytoma of the retina, or similar to schwannomas of the peripheral nerves. Complete removal was not possible. Gradual anesthesia of the entire 5th nerve distribution on the left developed. Preceding each advance of anesthesia, severe transitory pain was present, and paralysis of the left face developed. In the 10 months previous to surgery, 30 corneal ulcers of the left eye had occurred.

Bradley,2 in 1931, reported a neurocytoma of the Gasserian ganglion, the size of a cherry. The roots entering the ganglion were of normal size.

Cooper,5 in 1933, reported 3 cases of tumor of the Gasserian ganglion—a schwannoma or sheath neuroma, a neurocytoma, and a fibroblastoma.
The schwannoma was a discrete tumor eroding the petrous portion of the temporal bone. It measured $2 \times 1.5 \times 0.5$ cm. Following surgery, there was some defect in hearing on the left, residual pain in the left cheek and mouth, and palsy of the left 7th nerve. Seven weeks after operation there was complete left 5th, 7th, 9th, 10th and 12th nerve palsy, and bilateral involvement of the 8th nerve, more on the left side. The posterior fossa was explored by Dr. F. C. Grant and a tumor, $1 \times 5 \times 7$ mm. in size and situated directly over the foramen lacerum posterius, was removed. It had the same microscopical structure as the tumor removed from the middle fossa.

Cooper\(^5\) commented that clinical diagnosis is not always possible. Occasionally tic douloureux, atypical neuralgia, postzoster neuralgia, lues, tuberculosis, or an angle tumor may produce a clinical picture that cannot be distinguished from that of early tumor of the Gasserian ganglion. Exploration is justifiable in patients who suffer constant trigeminal pain not satisfactorily explained, and also in patients with objective indications of interruption of the trigeminal motor or sensory pathways without evidence of etiology other than ganglion tumor. Palliative section of the sensory root is justifiable when extirpation of the tumor is not feasible.

In 1932, Dandy\(^7\) found 18 tumors during 250 cerebellar explorations for trigeminal neuralgia. Unfortunately, he did not classify them. He stated that gross lesions, such as free arterial loops which lifted the sensory root from the brain stem, or venous branches crossing the nerve, were responsible for the trigeminal neuralgic pains in one-third of the cases.

Cohen,\(^5\) in 1933, tabulated all reported primary and secondary tumors of the Gasserian ganglion. Of 81 tumors, 22 were primary and 59 secondary. Thirty-three (and 1 questionable one) were exposed at surgery.

In 1935, Chang\(^8\) reported the successful removal of a neurofibroma of the Gasserian ganglion. Krayenbühl,\(^9\) in 1936, reported 2 cases, both neurofibromas. In one the patient died following a spinal puncture. Autopsy showed that the tumor arose in the sheath of the 5th nerve behind the ganglion, the bulk of the tumor being in the cerebellopontine angle on the left. The rostral part of the tumor arose in the Gasserian ganglion. He commented that the clinical features closely fitted those of an acoustic neurinoma. His second patient had a 6-year history of headache, sensory disturbance in the right side of the face, staggering, deafness, and mental deterioration. Ventriculography showed bilateral, symmetrical, internal hydrocephalus. No operation was performed. The patient died 2 years later. Autopsy revealed a tumor, $7 \times 4 \times 2.5$ cm., in the right cerebellopontine angle; it extended into Meckel's cave and beneath the floor of the 3rd ventricle. At the junction of the pons, the right 5th nerve was separated into two parts by the tumor. At its origin, the nerve was distinct from the tumor but in the distal part it was spread out diffusely and incorporated in the capsule of the tumor.

Krayenbühl,\(^9\) reviewed 54 cases of primary tumor of the Gasserian
ganglion, or root. In only 29 cases could the exact origin of the tumor be determined. In 26 cases, the tumor arose from the ganglion. He stated that the order in which symptoms appeared in these 54 cases made it possible to judge in every case whether the tumor originated in the middle or in the posterior fossa. He concluded that pain occurs when the ganglion is primarily involved and is usually absent when the tumor begins on the root. He also referred to the difficulty in distinguishing between tumor of the 5th cranial nerve root and an 8th nerve tumor. With tumor of the 8th nerve, the degree of deafness and vestibular loss is usually greater than the degree of 5th nerve sensory loss on the side of the lesion. With tumors involving the 5th nerve root, the degree of objective sensory loss is not always greater than the degree of deafness, so that precise differential diagnosis cannot be made. Their almost invariable forward extension into the middle fossa often produces ocular palsies. The presence of ocular palsies is a not uncommon finding with aneurysms of the internal carotid artery.

Cushing and Eisenhardt, in 1938, divided the Gasserian tumors into three principal groups: meningiomas, neurinomas, and tumors of malignant type.

In 1939, Fitzwilliams and Fell reported a case of metastasis in the Gasserian ganglion 1 1/2 years following radical breast surgery with subsequent implantation of radium needles. The tumor was of the scirrhous type. Autopsy revealed a basal meningitis with fluid pus on the meningeal surface. The brain was adherent near the right internal auditory meatus. No growths were found in the brain. A secondary carcinoma was present in the right Gasserian ganglion, the source of the meningitis.

Love and Woltman, in 1942, reported 2 cases with classical histories and signs of tic douloureux in which tumor of the ganglion was an incidental finding—a meningioma, and an epidermoid tumor. They mentioned a case of Adson's in which a meningioma of the right Gasserian ganglion was found during an operation for what was thought to be true tic douloureux. They stated that with tumors of the Gasserian ganglion pain is usually constant and often intense. It is not affected by eating, talking, shaving, or other external stimuli, as in tic douloureux. As a rule, paresthesia and sensory changes occur early. Shelden, according to Love and Woltman, pointed out that sensory changes may be lacking entirely, even when the ganglion has been almost completely destroyed, and that there may be disassociation of pain and temperature. This latter is also characteristic of involvement of the root of the 5th nerve. Usually paralysis of the motor root will be evident, following which the nerves in the region of the ganglion, especially the 4th cranial, will show involvement. The pain, so typical of tic douloureux, sometimes (but rarely) occurs with tumors of the 8th nerve, tumor of the pons, or multiple sclerosis. In the latter condition, the pain is often bilateral because of the central location of the lesion, and it may be relieved by section of the sensory root of the 5th nerve.

Alexander, in 1947, reported a case of central neurofibromatosis in
which nodular tumors involved both 5th nerves, the 6th and 8th on the right, while diffuse tumors of the right 3rd and 7th nerves were found. These were all neurofibromas. No cutaneous pigmented areas were present. He mentioned that polyneuritis cranialis must be considered with such diffuse intracranial involvement. This form of polyneuritis involves chiefly the 5th to the 12th cranial nerves in various combinations, and results from local disease—infecions, toxic, degenerative or neoplastic. It may accompany polyneuritis of the limbs.

CASE REPORT

TUMOR OF THE LEFT GASSERIAN GANGLION ASSOCIATED WITH MARKED ENLARGEMENT OF THE MANDIBULAR NERVE

J.W.S., a 37-year-old white male, was first seen on Mar. 13, 1951. Two years previously he had first noted a burning numbness in his left lower lip and chin, which gradually became more painful. The numbness continued, but the pain eased up for about a year. What pain he had was confined to the left lower lip and the distribution of the mental branch of the mandibular nerve. About September 1950, he began having pain in the left lower jaw. Four months later pain started in the tragus of his left ear and spread over his left cheek. There was soreness behind the left ear in the stylomastoid region. He also described pain on the anterior two-thirds of the left side of his tongue—a "kind of burning, like cigarette or tobacco bite." For the last 6 weeks he had had no freedom from discomfort in the left side of his face. He had also had some shooting pain from the left stylomastoid region to the angle of the left lower jaw. The pain did not go down the jaw, but at times he had pain in the region of his left lower lip and chin. Eating did not necessarily bring on the pain, although if he had pain, eating was troublesome. Shaving the

Figs. 1 and 2. Roentgenograms, Oct. 21, 1949. (1) Lower jaw, showing the dental canal of normal size on the left side. (2) Showing both mental foramina of equal size.
TUMORS OF THE GASSERIAN GANGLION

left side of his face was always painful. He thought there was some numbness of the left side of his face, compared to the right. He had had no pain in the left eye, left side of his nose or forehead, and his left nostril had been free of pain at all times. He had had no discomfort in the right side of his face.

Examination. The cranial nerves were entirely normal, with the exception of the left 5th. There was slight numbness of the left lower lip in the region supplied by the mental branch of the mandibular nerve. There seemed to be slight numbness of the tragus of the left ear, compared to the right. Otherwise, one could not outline any definite objective sensory changes. The corneal reflexes were active and equal and there was no anesthesia of the cornea. The motor component of both 5th nerves was intact and no evidence of wasting of the muscles of mastication was present. Both 7th nerves were normal. There were no other neurologic abnormalities.

Roentgenograms (lower jaw). Oct. 21, 1949 (Fig. 1): “The dental canal is of equal size on both sides of the mandible. The mental foramina (Fig. 2) are normal and equal in size.” Jan. 3, 1951 (Fig. 3): “There is extraordinary enlargement of the inferior dental canal and mental foramen (Fig. 4) on the left side. This enlargement apparently took place between Oct. 21, 1949 and Jan. 3, 1951. Enlargement of the mandibular nerve seems likely. The possibility of a tumor in this case must be considered, although its exact nature is not clear.” Mar. 26, 1951: “There is enlargement of the mandibular foramen on the left and the mental foramen on the left, and the canal within the bone which unites them is enlarged, as was shown on films of Jan. 3, 1951. The enlarged canal within the bone ends abruptly at the mental foramen and does not continue toward the symphysis. The vertical stereo shows a foramen ovale of normal dimensions on the right. On the left (Fig. 5) it is enlarged into a circular foramen of the same dimensions as the canal within the left mandible.”

Impression. All evidence pointed toward enlargement of the left mandibular
nerve, beginning as high as the Gasserian ganglion and extending throughout the entire length of the nerve to the mental foramen. This suggested von Recklinghausen's disease, but there were no other clinical manifestations of the disease.

Course. Exploration of the ganglion seemed justifiable. On April 17, 1951, biopsy of the mental nerve was reported as showing "marked inflammatory change and fibrosis in peripheral nerve (left mental)—etiology not apparent."

Reexamination, Aug. 16, 1951. The patient was not having so much pain but had noticed discomfort and numbness creeping up the left side of his face and check. He also thought there had been a little swelling along the lower jaw inside his mouth. His face was definitely swollen on the left. He had hypesthesia to pin-prick and touch in the distribution of the left mandibular division, and moderate hypesthesia in the distribution of the maxillary division, but no involvement of the ophthalmic division.

Roentgenograms, Aug. 17, 1951. The foramen ovale on the left side had increased slightly in size since Mar. 26, 1951. It was approximately three times the average size. The large mandibular canal on the left was again demonstrated. There was no definite change in the appearance of the mandible since the previous films.

Operation, Sept. 4, 1951. The usual approach to the Gasserian ganglion through the temporal route was made. After the middle meningeal artery was coagulated and cut, the dura was further dissected medially and a large reddish-gray tumor, 1.5 cm. in diameter, was encountered. It was globular in shape and comprised the enlarged Gasserian ganglion. Dissection was continued posteriorly until the tumor was completely mobilized and free. The sensory root of the 5th nerve was sectioned posterior to the ganglion. The motor root was not identified. The tumor was teased forward, freeing it from the wall of the cavernous sinus. The tumor extended into the 2nd and 3rd divisions, but did not seem to invade the 1st division grossly. All three divisions were cut and the tumor was then removed in three pieces. The foramen ovale was about three times the normal size. The tumor tissue that extended into the foramen rotundum and ovale was cauterized. The distal portion of the ophthalmic division, which appeared grossly normal, was not cauterized for fear of damage to the nearby oculomotor and trochlear nerves.

Pathologic Report (Dr. L. J. Tragerman). "There is a large coarse nodular structure suggesting possible pre-existing bundles of fibres. These nodules are occupied by cells of two principal appearances. One is in the form of solid masses, with a tendency to gland-like arrangement, but without glandular formation. The individual cells are characterized by distinct cell borders, pale to coarsely granular cytoplasm and large pale nuclei. Nucleoli are prominent. The other cell-type pattern is that of a spindle-cell proliferation. One sees the preponderance of large pale cells
TUMORS OF THE GASSERIAN GANGLION

resembling epithelial cells. In some areas the cells are spindle in type, with elongated oval nuclei. The tumor appears histologically malignant. Diagnosis: Probable malignant schwannoma.”

Pathologic Report (Dr. Cyril B. Courville). “Nine slides were studied which were stained with hematoxylin and eosin, phosphotungstic acid hematoxylin, and Perdrau stains. This appears to be a tumor which is pressing into the nerve. The architecture is most unusual but suggestive of a neurofibroma.”

Final Diagnosis. Unclassified tumor which resembles a neurofibroma (Fig. 6).

Fig. 6. Photomicrograph of tumor of the Gasserian ganglion showing the unusual architecture suggesting probable malignant schwannoma or neurofibroma.

Reexamination, Dec. 31, 1951. The patient had noted intermittent swelling in the region of the left temple. At times he had had twinges of pain above the left zygoma, upper eyelid, and in the low frontal region. He had some very slight discomfort most of the time, and occasionally sharp transitory pain. There was anesthesia in the entire left trigeminal distribution. It was less marked in the 1st division, and corneal sensation was retained. There was slight motor weakness of the left 5th nerve, the jaw deviating to the left.

COMMENT

Tumors of the Gasserian ganglion may be either primary or secondary. In 1933, Cohen reported a total of 81 tumors of the ganglion, of which only 22 were primary, and 59 were secondary. Apparently endothelioma is the most common type of primary tumor, followed by fibrosarcoma, and neurofibroma, other types being extremely rare.
It is possible in some cases to determine the origin of the tumor (whether in the middle or posterior fossa) when the trigeminal nerve is involved. The order in which symptoms appear is of great importance. Pain occurs when the Gasserian ganglion is primarily involved and is usually absent when the tumor begins on the sensory root. The onset of pain in the distribution of the 5th nerve, which later decreases or disappears, should make one suspect extension of the tumor from the ganglion along the sensory root into the posterior fossa. Lower cranial nerve palsies, cerebellar or cerebral peduncle signs should point to involvement of the posterior fossa by the tumor. It may be very difficult to distinguish between tumor of the 8th nerve (acoustic neurinoma) and tumor of the 5th cranial nerve root. The degree of deafness with 8th nerve tumor and vestibular loss is greater than the degree of 5th nerve sensory loss on the side of the lesion. In tumors involving the 5th nerve root, the degree of objective sensory loss is not always greater than the degree of deafness, so that precise differential diagnosis cannot be made. Sensory changes may be lacking entirely, even when the ganglion has been almost completely destroyed. There may be dissociation of pain and temperature, this latter being also characteristic of involvement of the 5th nerve root. Usually paralysis of the motor root can be demonstrated, and at times involvement of the 3rd and 4th cranial nerves, which are in close proximity to the ganglion, will appear in the presence of a tumor of the ganglion. The pain, so typical of tic douloureux, may on rare occasions occur with tumors of the 8th nerve, tumor of the pons, atypical neuralgia, postzoster neuralgia, lues, tuberculosis, and multiple sclerosis. In the latter case the pain is often bilateral.

It was indeed fortunate that roentgenograms of the mandible were made at the onset of this patient's symptoms, which could be compared with subsequent views. These show the rapidity with which this tumor had invaded the entire mandibular canal, including the mental foramen, between Oct. 21, 1949 and Jan. 3, 1951. One cannot help but wonder if similar lesions may have sometimes escaped detection and been classified as atypical neuralgia, because of the few objective clinical findings, and the peculiar nature of the subjective complaints.

CONCLUSION

An unusual case of tumor involving the Gasserian ganglion and the mandibular and maxillary divisions of the trigeminal nerve is reported. One can only speculate as to whether the tumor arose primarily in the ganglion and subsequently spread along the entire mandibular division, later involving the maxillary division, or arose primarily in the mandibular division, and later involved the Gasserian ganglion. The diagnosis of probable malignant schwannoma was made by one pathologist, that of an unclassified tumor resembling a neurofibroma by another. From the appearance of the enlarged mandibular canal, without further invasion of the mandible, it would appear that this was a rapidly growing primary tumor.
of the mandibular nerve, a malignant schwannoma, which involved the Gasserian ganglion.

REFERENCES


