Detection of small trigeminal neurinomas

ROBERT LEVINTHAL, M.D., AND JOHN R. BENTSON, M.D.

Department of Surgery (Neurosurgery) and Radiological Sciences, UCLA School of Medicine, Los Angeles, California

Four cases of trigeminal neurinoma are reviewed with particular attention to clinical signs and symptoms, lumbar puncture, electroencephalogram, brain scan, plain skull and tomographic radiographs, and angiographic and pneumoencephalographic findings. Pneumoencephalography, with special tomographic projections to identify various portions of the trigeminal nerve, delineated the tumor in all cases. Tumor removal was complete in three patients and nearly complete in the fourth. There was no operative morbidity or mortality.

KEY WORDS  •  trigeminal neurinoma  •  pneumoencephalography

TRIGEMINAL neurinomas are relatively rare tumors that arise from the sheath of Schwann and constitute 2.9% of intracranial neurinomas and 0.26% of all brain tumors. They are well circumscribed, soft, relatively avascular tumors with a thin capsule.

Since they are slow-growing, benign tumors, they often attain a large size before being detected clinically and radiographically. To remove these tumors without mortality or severe morbidity they must be discovered when they are relatively small and not intimately attached to the adjacent brain. A review of four patients with trigeminal neurinoma seen between 1971 and 1974 at the UCLA Hospital yielded clinical and radiographic clues that could facilitate early detection of these tumors.

Case Reports

Case 1

This 57-year-old man had a 10-year history of decreased hearing on the left and a 2-year history of tingling in the left cheek. One year prior to admission, he noted left retroorbital pain, described as a feeling that his "left eye was on fire." He also noted decreasing bulk of the left side of his face.

Examination. Neurological examination disclosed decreased bulk of the muscles of mastication on the left, decreased left corneal response, and left facial hypalgesia and hypesthesia, greater in the second and third division of the fifth cranial nerve (V2 and V3). Lumbar puncture showed normal pressure and a cerebrospinal fluid (CSF) protein of 34 mg%. Audiometric examination revealed high-frequency hearing loss bilaterally with Type I Békésy audiometry, high short increment sensitivity test (SISI), and no tone decay. Electroencephalogram and isotope brain scan were within normal limits. Plain skull films showed erosion of the left petrous apex. Tomograms of the base of the skull demonstrated a smooth-walled erosion of the floor of the left middle fossa (Fig. 1). Bilateral carotid and left vertebral angiograms were within normal limits. The pneumoencephalogram showed slight elevation of the left temporal horn and the left cerebral peduncle. No mass was visible in the cerebellopontine angle, and the pons was not tilted.
Trigeminal neurinomas

Operation. A left temporal craniectomy was performed and an extradural approach used to expose a dark, yellow-purple tumor that involved the Gasserian ganglion and extended into the foramen ovale. The tumor, 2½ cm in diameter, was removed in piecemeal fashion. The pathological diagnosis was trigeminal neuroma.

Postoperative Course. The patient did well and soon returned to full employment. Examination 6 months postoperatively demonstrated hypalgesia and hypesthesia of the left side of the face, slightly decreased corneal reflex on the left, and deviation of the jaw to the left.

Case 2

This 56-year-old right-handed woman suffered a "whiplash" injury in a minor automobile accident 3 years prior to admission. Approximately 1 month after the accident, she noted right-sided headaches with right facial pains that radiated from the right temporal area to below the right eye. Skull and cervical spine films, brain scan, and electroencephalogram performed 2 months prior to her admission to the UCLA Hospital were within normal limits. She was treated with carbamazepine (Tegretol), with slight relief.

Examination. On admission she had unsustained nystagmus on right lateral gaze, decreased corneal reflex on the right, minimal ptosis of the right eye, and a questionable Babinski sign on the left. Her facial sensation was within normal limits. Lumbar puncture was unremarkable except for a CSF protein of 64 mg%. Audiometric examination was within normal limits. Right internal and external carotid angiograms were negative. Attempts to catheterize the tortuous vertebral arteries were unsuccessful. Pneumoencephalogram (Fig. 2) with posterior fossa and temporal horn tomograms demonstrated a mass extending posteriorly over the right petrous apex, elevating the right cerebral peduncle. There was also elevation of the right temporal horn.

Fig. 1. Case 1. Basal skull tomography demonstrates enlargement of the left foramen ovale.

Fig. 2. Case 2. Right trigeminal tumor occupying the middle and posterior fossae. Left: Pneumoencephalogram. Frontal tomogram through midbrain demonstrates elevation of the right cerebral peduncle by underlying mass (arrows). Right: Tomogram of the petrous apices shows the normal left fifth nerve, seen end-on as a round density (solid arrow). The fifth nerve tumor on the right is capped by a thin strip of cisternal gas (open arrows).
R. Levinthal and J. R. Bentson

Fig. 3. Case 3. Comparison of angiographic and pneumoencephalographic findings of a right fifth nerve tumor. Left: Vertebral angiogram, Towne's projection, demonstrates elevation of the right superior cerebellar artery (arrow). Right: Vertebral angiogram, venous phase. There is a subtle elevation of the right cerebral peduncle relative to the left (small arrows) and lack of filling of the right petrosal vein compared to the left (open arrow).

Operation. A right temporal craniotomy with intradural approach revealed a tumor, 2 × 1.5 cm in size, lying within Meckel’s cave. The tumor was totally resected.

Postoperative Course. The patient had a prolonged postoperative recovery period complicated by seizures that were successfully treated with diphenylhydantoin. Two months postoperatively she returned to her normal activities. She then demonstrated weakness of the fourth nerve on the right side (diplopia on left lateral, downward gaze), decreased corneal reflex on the right, and hypalgesia and hypesthesia of the right V1 and V2 distributions.

Case 3

This 55-year-old right-handed woman was well until 4 months prior to admission, when a right facial numbness began at the angle of the mouth and progressed over a few weeks to involve the entire right side of the face, including the forehead, tongue, and oral mucosa. Approximately 1 month prior to admission, she experienced the sudden onset of lancinating pains involving the entire right side of her face and lasting approximately 20 minutes; the pain responded to mild analgesics. She had also experienced transient sharp, repetitive dysthesias in the right suprorbital region. Two months before admission, she was admitted to another hospital where skull films, electroencephalogram, and isotope brain scan were within normal limits.

Examination. Neurological examination revealed hypalgesia and hypesthesia of the right V1, V2, and V3, decreased right corneal reflex, and right masseter weakness. Laboratory examinations were unremarkable except for CSF protein of 101 mg%. Electromyogram of facial muscles showed denervation of the right masseter and temporalis muscles. Audiometric examination was normal. Skull films, skull base tomograms, sinus films, isotope brain scan, and electroencephalogram were unremarkable.

Angiogram revealed asymmetry of the superior cerebellar arteries, with the right higher than the left (Fig. 3 left). The right petrosal vein did not fill (Fig. 3 right). The right cerebral peduncle was slightly higher than the left, and there was some elevation of the right transverse pontine vein.

The pneumoencephalogram with tomograms of the posterior fossa demonstrated some flattening of the right pons and swelling of the root of the right fifth nerve. There was only partial filling of the right cerebellopontine angle cistern. Tomograms of the petrous tips showed that the fifth nerve was normal on the left, but that there was a mass extending through the tentorial notch on the right. The temporal horns were in the normal position.

Operation. A right temporal craniotomy was performed. A trigeminal neurinoma dis-
Trigeminal neurinomas

placing the brain stem (as described above) and with only minimal extension into Meckel's cave was totally resected.

Postoperative Course. Postoperatively, a right fourth nerve palsy was noted, but this resolved. A herpetic lesion developed in the right V3 distribution. The patient was discharged on the 26th postoperative day; the only deficit remaining was a right fifth nerve palsy.

Case 4

This 61-year-old right-handed woman experienced numbness and pain of the left cheek and jaw 1 year prior to admission. Over a period of 1 week, the numbness spread to include the forehead and anterior scalp. She noted that it became progressively more difficult to chew on the left side and that she would bite her left cheek and tongue. Six months prior to admission, she had noted redness and excessive tearing of her left eye. Three months before admission, she felt "stuffiness" of the left ear. She complained of intermittent double vision, and on one examination by her private physician, a left sixth nerve weakness was demonstrated.

Examination. Neurological examination revealed a fine horizontal and vertical nystagmus, the left pupil ½ mm larger than the right, a left sixth nerve weakness, left tongue weakness and atrophy, hypesthesia and hypalgesia of all divisions of the left fifth nerve, decreased corneal reflex on the left, and deviation of the jaw toward the left. Examination of CSF was normal, and cytological tests were negative.

Plain skull films and skull tomograms demonstrated erosion of the medial aspect of the left petrous pyramid and the floor of the left middle fossa. A brain scan was normal. Electroencephalogram demonstrated occasional sharp waves and asymmetrical slowing in the left temporal region.

Bilateral internal and external carotid angiograms were normal. Left vertebral angiogram showed tilting of the cerebral peduncles and the pons on the half-axial films at the venous phase. The pneumoencephalogram with posterior fossa and temporal horn tomograms showed a mass extending posteriorly from the area of erosion of the left petrous tip into the left cerebellopontine angle (Fig. 4). There was distinct elevation of the left cerebral peduncle. The left temporal horn was very slightly elevated.

FIG. 4. Case 4. The tumor lying at the origin of the left fifth nerve fills the left angle cistern (arrows).

Operation. A left temporal craniotomy with extradural approach revealed a soft, yellow neurinoma originating from the Gasserian ganglion with extension into the foramen ovale and rotundum and into the pterygoid region. The tumor extended along the fifth nerve root into the posterior fossa. The tentorium was divided to expose and remove the infratentorial portion of the tumor. Resection was complete except for a small rim of tumor along the cavernous sinus.

Postoperative Course. The patient did well postoperatively. She had double vision secondary to a mild left fourth nerve palsy, analgesia in the distribution of the left fifth nerve, and an absent left corneal reflex. Her masseters were equal in strength. A transient cerebrospinal rhinorrhea resolved with frequent lumbar punctures and conservative management.

Discussion

Trigeminal neurinomas frequently originate within Meckel's cave in the middle cranial fossa and may extend along the trigeminal root to involve both the middle and posterior cranial fossae. They may, however, arise independently from the trigeminal root and involve only the posterior cranial fossa. Since 1935, 57 trigeminal neurinomas, verified surgically or pathologically, have been described in the English literature. Of those cases in which the loca-
tion was described, 24 were located mainly within the middle cranial fossa, 15 were located mainly within the posterior cranial fossa, and 13 involved both fossae equally. In our series of four cases, two tumors were found mainly in the middle fossa, one was located mainly in the posterior fossa, and one shared both cranial fossae equally.

Jefferson characterized the topography of these lesions in the following statement: "Let it be said at once that practically all the tumors which presented in the middle or posterior fossa were either known to have or could have an extension into the other fossa . . . the gross form of these tumors is almost always that of an hour-glass but with a large end and a small end, joined together by a short thick neck that erodes the superior petrosal apex."

The clinical presentation of trigeminal neurinomas is protean. The symptom complex generally depends on the anatomical location of the bulk of the tumor. A middle-fossa tumor characteristically is associated with evidence of fifth nerve sensory involvement. Irritation or sensory impairment of the fifth cranial nerve is the presenting symptom in more than one-half of the reported cases. However, it is not unusual to find a verified trigeminal neurinoma with no fifth nerve symptoms. It has been suggested that tumors arising from the trigeminal ganglion are often associated with pain, whereas those arising from the root are not. Jefferson believed that pronounced facial wasting and numbness was uncommon with a benign lesion. Other findings in middle-fossa syndromes are related to the size of the tumor and the presence of increased intracranial pressure. Posterior-fossa symptoms are presenting complaints in approximately one-fourth of trigeminal neurinomas and are eventually present in about one-half of the cases. The most common symptoms are nystagmus, facial weakness, and ataxia.

Laboratory examinations often reveal an increase in CSF protein concentrations. Brain scans are helpful, provided that the tumor is sufficiently large and in a favorable location, and electroencephalograms may occasionally be of localizing significance in middle fossa lesions.

Our four patients all had evidence of fifth nerve dysfunction, irritation, or both. The presence or absence of pain was not helpful in localizing the tumor to the root or ganglion. There was a paucity of other signs and symptoms, probably related to the relatively small tumor size and lack of intracranial pressure. Two cases had increased CSF protein (64 and 101 mg%). Only one electroencephalogram showed a focal abnormality, temporal slowing. All four patients had normal brain scans.

The importance of radiographic signs in the diagnosis of trigeminal neurinomas has been stressed in nearly all communications. Early emphasis was placed on skull-film changes, which continue to be the most characteristic radiographic signs of these tumors. Lindgren described in considerable detail the erosion of the base of the middle fossa with widening or loss of the foramen ovale and noted that destruction of the tip of the petrous pyramid was a sign that the tumor also entered the posterior fossa. There has been mention in the literature of the value of tomography, which we find to be the most helpful in anteroposterior and basal projections.

Early reports dealt with very large tumors, as evidenced by frequent mention of sellar erosions. More recently, as smaller tumors have been diagnosed, the percentage of positive skull findings has decreased. Knudsen and Kolze found no skull-film abnormalities in four patients seen between 1965 and 1970. Of our cases, only the two largest tumors, Cases 1 (Fig. 2) and 4, both primarily located in the middle fossa, had the typical skull erosions. The other two had no skull defects recognizable on extensive plain skull film and tomographic studies.

Arteriography was considered to be of little value in early reports except to exclude aneurysms. Westberg reviewed 10 patients with trigeminal neurinomas on whom carotid angiography had been performed and noted carotid displacement in six. In these cases, the extradural segment of the internal carotid artery was displaced forward, downward, and medially. Enlarged cavernous carotid branches related to the tumor were found in the majority of his cases. Similar reports of angiographic findings have been made by several other authors. Carotid arteriograms were done bilaterally in our four cases, and careful comparison of the two sides showed no carotid displacements related to the tumors. Subtraction prints showed neither vascular staining nor unusually prominent
Trigeminal neurinomas

Fig. 5. Upper Left: Diagram illustrating the planes of tomography used to demonstrate the trigeminal nerves. Upper Right: Pneumoencephalographic tomogram using the projection plane "A" on the diagram. This projection shows the fifth nerves as round or oval densities within the petrous notches (arrows). Lower Right: Pneumoencephalic tomogram using the projection plane "B" of the diagram. The fifth nerves are seen as vertical bands within the cerebellopontine angle cisterns (arrows).

Meningeal vessels. Vertebral arteriograms, on the other hand, were positive in two of the three cases in which they were performed. The principal findings were in the venous-phase films that demonstrated asymmetry of the peduncles and tilting of the pons in two and poor filling of the ipsilateral petrosal vein in one (Fig. 3). These findings mirrored the pneumoencephalographic changes in the same patients but were more difficult to appreciate.

The pneumoencephalographic findings were the most impressive in our cases, and we wish to emphasize their importance. Temporal horn elevation was apparent in two cases, a subtle finding in the third case, and not present in the fourth. Two points should be kept in mind in evaluating subtle differences in position of temporal horns: first, both horns must be completely filled on the same non-rotated film; and second, asymmetry in the heights of the petrous ridges will cause apparent asymmetry of the temporal horns. In our experience, the distance between each petrous ridge and the temporal horn at that level, measured on a tomogram, is remarkably similar, with differences of 2 mm or more indicating pathological asymmetry of the temporal horns. In all four cases, the tumor was demonstrated on the pneumoencephalographic tomograms at the level of the petrous apex, although bone destruction of the apex was only present in two.

We used a special projection for filming the fifth nerve at the petrous apex (Fig. 5). After injection of gas into the ventral basal cisterns by extending the head of the seated patient, the pneumoencephalographic chair was rapidly moved to the supine position. A series
of coronal tomograms was then obtained in the region of the petrous apices. The plane of the tomogram is approximately at a right angle to the canthomeatal line. Normally, the fifth nerves are seen distinctly as small round densities adjacent to the petrous notches. In these cases of trigeminal neurinomas, a mass was seen in that location, and the ipsilateral cerebral peduncle was shown to be elevated in each case. Prior to the filming of the tomographic series just described, a series of coronal tomograms had been obtained of the brain stem of the seated patient whose head and neck were well flexed forward. These tomograms, taken routinely whenever posterior-fossa masses are expected, normally will demonstrate the origins of the fifth nerves from the pons and their courses toward the petrous apices (Fig. 5). When the fifth nerve tumor extends to the exit of the nerve from the pons, one sees a mass adjacent to the pons (Fig. 3) or lack of filling of the cerebellopontine angle cistern (Fig. 4). This projection is not as satisfactory in demonstrating the fifth nerves passing the petrous apices as the previously described projection. The tumor was demonstrated in each of the four cases of this series by tomography at the petrous apex and midbrain levels, while tomograms at the cerebellopontine angle level showed the tumor in two cases. Both sets of tomograms should be obtained, for they will give valuable information regarding the extent of spread of the tumor into the posterior fossa.

Schisano and Olivecrona reviewed the literature and reported an overall operative mortality rate of 41%. However, in their personal series of 19 operated cases, they totally removed the tumor in eight cases, almost totally in four, and partially in seven; they had only one operative death, and two tumor recurrences. Knudsen and Kolze had one operative mortality in their four cases. Paillas, et al., stressed that total tumor removal should be attempted but that a high operative mortality could be expected; they had 37.5% mortality in their cases. For total tumor removal without morbidity, the tumor must be discovered before it has reached a size sufficient to cause increased intracranial pressure or involve vital structures. In our series, total tumor removal was accomplished in all but one case in which a small rim of tumor was left along the border of the cavernous sinus. There were no deaths. Transient worsening of fifth nerve function took place in all patients, but all returned to preoperative levels or better. Two patients also demonstrated transient diplopia secondary to fourth nerve weakness, probably from manipulation about the tentorium. One patient had cerebrospinal rhinorrhea that resolved. Another had one generalized seizure but has had no recurrence since being placed on diphenylhydantoin.

Conclusions

Although recognizable syndromes are associated with trigeminal neurinomas, it may be difficult in early cases to determine clinically those patients with facial pain and numbness who have trigeminal neurinomas. Radiographic examinations are, therefore, of considerable importance in the detection of these tumors. The classical bone erosions are nearly specific for fifth nerve tumors but cannot be expected to be present in the majority of cases unless there is a long delay between onset of symptoms and radiographic examination. The same is true of angiography. The accuracy of computer axial tomography in the detection of small benign tumors adjacent to the skull base has so far been disappointing. Pneumoencephalography is the most reliable means of detecting these tumors. Close attention must be given to the positions of the temporal horns and to tracing each fifth nerve from its origin to the point where it passes the petrous apex. A special projection useful for detecting the tumor at the petrous apex is described.

With these pneumoencephalographic techniques, the presence of a trigeminal neurinoma was convincingly demonstrated in our four cases, whereas bone changes were found in only two. Angiography was also positive in two cases but demonstrated tilting of the brain stem rather than the changes of the pre-cavernous internal carotid artery which had been previously described. Fifth nerve symptoms of irritation or dysfunction, even if associated with unremarkable skull films, tomograms, isotope brain scan, and electroencephalogram, should be vigorously pursued by angiography and particularly by pneumoencephalography with tomography.

References

Trigeminal neurinomas


Address reprint requests to: Robert Levinthal, M.D., Department of Neurosurgery, UCLA School of Medicine, Los Angeles, California 90024.