Trigeminal neurinoma in adolescents

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Neurinomas comprise only a small percentage of all intracranial tumours and among them acoustic neurinomas are the most common. However, those originating from the trigeminal ganglion or nerve root, seventh nerve, or other lower cranial nerves are considered rare. Intracranial neurinomas have appeared in the literature under various nomenclature, such as neurofibroma, perineurial fibroblastoma, neurilemmoma, and Schwannoma (Russell and Rubinstein, 1963; Zülch, 1965).

Reviews on trigeminal neurinomas were reported by Loew and Tönnis (1954), Jefferson (1955), Olive and Svien (1957), Schisano and Olivecrona (1960), and in other case reports (Altmann, 1928; Nowotny and Uiberall, 1934; Glaser, 1935; Krayenbühl, 1936; Shutova, 1940; Gordy, 1965; Best, 1968). About 95 cases of trigeminal neurinomas are known in the literature, of which only eight were found in adolescents.

We would like to add two cases of trigeminal neurinomas occurring in adolescents, stressing the radiological features and the surgical approach.

CASE 1

A 14-year-old boy was admitted to the hospital in January 1966 with a two-year history of progressive numbness of the left side of his face. He also experienced sharp pains and blurring of vision of his left eye and occasional double vision on left lateral gaze. Six months before admission, an unsteady gait developed with a tendency to fall to the left. Other complaints included tinnitus in his left ear, and difficulty with chewing and swallowing.

Abnormal findings on neurological examination were limited to the left side: a slight blurring of the optic disc, mild ptosis, sixth nerve paresis, and a mild facial weakness. Facial sensation was impaired with some sparing in the mandibular division and absent corneal sensation. There was marked atrophy and weakness of the masticatory muscles with deviation of the jaw. Vestibular testing resulted in a hyperactive response on the left, but hearing was intact. There was no dysphasia, but his speech

was garbled. Deep tendon reflexes were depressed in the left upper limb, but were equal in the lower limbs with bilateral extensor plantar reflexes and unsustained ankle clonus. Cerebellar testing revealed dysmetria and dysidiadochokinesia, more prominent on the left. Mild weakness of the left arm was present. He walked slightly broad-based and veered to the left.

Electroencephalogram showed moderately severe slow wave abnormality and a few sharp waves in the left temporal area. Skull films revealed thinning of the posterior clinoid process and, in the submento-vertex view, a large osteolytic defect involving the petrous apex extending into the middle fossa and greater sphenoid wing. Erosion of the petrous apex appeared sharply circumscribed and the foramen ovale was completely lost (Fig. 1). On vertebral angiography, the basilar artery was displaced posteriorly and the left superior cerebellar and posterior cerebral arteries were notably elevated and in the lateral view were bowed upwards (Fig. 2). In the lateral carotid arteriogram, the extradural part of the siphon appeared straightened and displaced forward and the distal siphon was stretched and opened, due to elevation of this segment and of the proximal middle cerebral artery. The anterior choroidal artery was similarly stretched and elevated. In the anteroposterior view, the siphon was displaced medially and opened with elevation of the middle cerebral artery. The anterior cerebral artery was bowed to the right (Fig. 3). In the lateral phlebogram, the basal vein was notably elevated and abnormal vessels were identified in the para- and retrosellar areas (Fig. 4). With these clinical and radiographic findings, a neurinoma of the trigeminal nerve was suspected.

Through a left temporal osteoplastic craniotomy, the temporal lobe was elevated intradurally. At the medial aspect of the middle fossa an encapsulated tumour was encountered, extending anteriorly to the temporal tip and posteriorly through the petrous erosion and underneath the tentorium into the posterior fossa. An intracapsular tumour removal was accomplished and the greater part of the tumour capsule removed, save for the medial wall densely adherent to the brain-stem and to the cavernous sinus. At the floor of the middle fossa the flattened gasserian ganglion and the thinned third division of the trigeminal nerve were seen.

HISTOLOGY The tumour had the characteristic structure of a neurinoma composed of mature nerve fibres and

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FIG. 1. Submento-vertical view of the skull shows a large osteolytic defect in the floor of the left middle fossa and a sharply circumscribed erosion of the apex of the petrous bone. The foramen ovale on the left is completely lost.

Schwann cells following the Antoni A and B patterns. The tumour was well vascularized with no suggestion of malignancy.

Post-operatively, in addition to the deficit of the left trigeminal nerve, the fourth and sixth nerves were also paretic. In the course of several months, the sixth nerve palsy recovered completely and diminished sensation was felt over the third division. Speech, co-ordination, and gait were normal and corrective eye surgery was carried out in July 1967.

He was readmitted in April 1968 with complaints of intermittent headache of six weeks' duration, recurrence of slurred speech, unsteady gait, and fatigue. Examination revealed the left trigeminal nerve deficit unchanged and, in addition, a mild facial weakness and diminished hearing. The same reflex changes, cerebellar signs, and ataxic gait were present again.

Electroencephalogram was unchanged and an Hg^{197} brain scan was negative. Cerebrospinal fluid protein was 86 mg/100 ml. Carotid and vertebral arteriograms were repeated and a pneumoencephalogram was carried out. The latter showed posterior displacement of the aqueduct and fourth ventricle, and only little shift of the structures to the right, consistent with tumour recurrence in the posterior fossa.

The previous temporal craniotomy was reopened and the temporal fossa found void of tumour. After incising the tentorium, the plum-sized tumour was located anteriorly to the cerebellum and medially along the brain-stem and within the petrous bone. An incomplete intracapsular tumour removal was accomplished. Histologically, the tumour again showed characteristics of a neurinoma, but no evidence of malignancy.

FIG. 2. Left vertebral arteriogram. The basilar artery is slightly displaced posteriorly. The left superior cerebellar and posterior cerebral arteries proximally are notably elevated and in the lateral view are convex upwards.

FIG. 3. Left carotid arteriogram. The carotid siphon is displaced anteriorly and medially and is stretched in its distal portion. The proximal middle cerebral artery is elevated and the anterior choroidal artery is similarly stretched and elevated. The anterior cerebral artery is slightly bowed to the right.

FIG. 4. In the lateral phlebogram, the basal vein is rather notably elevated well forward to the level of the anterior clinoid. Abnormal tumour vessels are identified in the parasellar and retrosellar areas.

Follow-up examination one year later showed the trigeminal nerve deficit unchanged and nystagmus on upward and right lateral gaze. Gait was still slightly broad based and there was minor dysmetria and slight weakness of the left hand.

CASE 2

Two years before admission, this 16-year-old boy developed a 'white speck' over the cornea of his right eye, of

which he was not aware. A painless inflammation of the eye developed and was treated with eye drops and ointments. Six months later, his relatives noticed that the patient was unable to walk in a straight line. Weakness of the right hand developed and progressed to involve the right side. This was followed by dysarthria. On direct questioning, the patient admitted having occasional frontal headaches for one year, as well as diminution of smell and hearing on the right side.

On examination, the patient was a poorly nourished, conscious, co-operative, young boy, who was grossly dysarthric. His gait was ataxic. The loss of smell was attributed to allergic rhinitis. A dense corneal opacity on the right side prevented examination of the fundus and the left fundus was normal. The right fifth (both sensory and motor function), sixth, seventh, eighth, ninth, and tenth cranial nerves were paralysed and he was unable to protrude his tongue. There was right-sided hemiparesis with gross cerebellar signs in the form of bilateral horizontal nystagmus, finger-nose and heel-knee ataxia, hypotonia, and depressed tendon reflexes. A clinical diagnosis of cerebello-pontine angle tumour was entertained.

Plain skull films revealed enlargement of the pituitary fossa, sutural diastasis, and gross destruction of the apex of the right petrous bone. A right brachial arteriogram outlined both the right carotid and vertebrobasilar systems. The basilar artery was stretched and grossly displaced to the left. The posterior cerebral artery was elevated. The carotid siphon was markedly unfolded and grossly displaced and the middle cerebral artery was also elevated. Pathological vessels were seen in the region of the apex of the petrous bone and extending into the middle cranial fossa (Fig. 5).

FIG. 5. Case 2. Brachial arteriogram shows the gross displacement of the carotid siphon and major cerebral vessels and some abnormal vessels in the area of the tumour. In the AP view the destruction of the right petrous apex is evident.

A right temporal osteoplastic craniotomy was performed under controlled hypotension and hypothermia. After opening the dura, the temporal lobe was retracted and it became apparent that the tumour in the middle fossa was extradural. The free edge of the tentorium was visualized and incised, extending the incision laterally along the superior border of the petrous bone. In the region of the cavum of Retzius, two layers of dura had to be incised separately before exposing the tumour. The tumour filled the whole middle fossa and extended into the cerebello-pontine angle. Starting in the middle fossa, the tumour was initially removed piecemeal. Having reduced its size, the remaining tumour was dissected from the surrounding structures and removed en masse, amputating it at its junction with the infratentorial part. The opening in the tentorium was enlarged and the tumour in the cerebello-pontine angle was totally excised in large pieces. In the middle fossa some of the tumour capsule was left medially for fear of damaging the cavernous sinus. The fifth nerve was intimately involved in the tumour. During excision of the tumour, the third nerve was seen medially while the sixth, seventh, and eighth nerves were in its bed and the ninth and tenth nerves were stretched at the lower pole. Venous bleeding along the superior petrosal sinus was troublesome, but finally controlled. The opening in the tentorium was not closed. The cerebrospinal fluid was flowing freely into the tumour cavity.

The post-operative period was uneventful. At the time of discharge, the right fifth and sixth nerves were completely paralysed, but the rest of the cranial nerves were showing signs of recovery. The cerebellar signs and right hemiparesis were improving.

On follow-up examination 10 months later, the only positive findings were a complete right fifth and sixth nerve palsy and a minimal degree of ataxia.

DISCUSSION

In Zülch's series of 4,000 brain tumours, 7.5% were neurinomas (1965). In the report by Nowotny and Uiberall (1934), the occurrence of primary tumours in the trigeminal area was quoted as 1 to 1.5%, neurinomas representing one-fourth of them. Schisano and Olivecrona (1960), in their series of 5,727 verified brain tumours, found 442 neurinomas (7.7%) and, of these, only 15 originated from the ganglion or posterior root of the trigeminal nerve, thus comprising 0.2% of all tumours. They also pointed out that earlier studies on tumours in this area were proven not to be neurinomas but either other benign or malignant tumours. Of the 95 verified trigeminal neurinomas quoted in the literature, and the two cases reported, 10 occurred in adolescents. This represents a considerable incidence, since meningiomas and neurinomas are usually not associated with childhood and adolescence.

The histological appearance in the two cases reported was typical of a neurinoma and showed no evidence of malignancy in spite of the early recurrence of the tumour in case 1. Trigeminal neurinomas originate from the sheath cells of the

nerve fibres and may either develop from the gasserian ganglion or the root of the trigeminus. The majority of the described tumours originated from the gasserian ganglion and were located in the middle cranial fossa in comparison with the few in the posterior fossa originating from the trigeminal root. However, many of the large tumours from the ganglion presented as 'hourglass-shaped tumours' in both cranial fossae. Of the 10 adolescent patients, the tumour originated from the gasserian ganglion in eight and posterior fossa symptoms were present in six, indicating extension of the tumour (Table I).

The symptoms are, characteristically, sensory changes and motor weakness of the involved trigeminal nerve. Facial pain has not been a significant feature. Paresis of the masticatory muscles appears later in contrast with the early appearance of sensory changes. The optic pathways may be involved, but papilloedema is not a frequent occurrence. According to the size of the tumour, other cranial nerves, especially the sixth, seventh, and eighth may be affected. With extension into the cerebello-pontine angle, cerebellar and pyramidal signs are present frequently. Several authors postulated that tumours arising from the nerve root are not associated with facial pain but cause sensory changes in the face and that the symptom of facial pain is more characteristic of tumours of the gasserian ganglion (Glaser, 1935; Krayenbühl, 1936;

Schisano and Olivecrona, 1960). However, these rulings are not constant findings, especially with extensive tumours.

Some of the radiographic features seen with trigeminal neurinoma are quite characteristic and have been pointed out in earlier reports (Altmann, 1928; Krayenbühl, 1936; Taveras and Wood, 1964; Best, 1968). The most common skull film change is widening of the foramen ovale which may extend into a bony defect of the middle fossa. With large tumours, a soft tissue mass may be seen in the pterygoid fossa and posterior nasopharynx. Extension of the tumour into the posterior fossa is always associated with erosion of the petrous apex. Holman, Olive, and Svien (1961) pointed out that the petrous erosion medial to the internal acoustic meatus with trigeminal neurinomas is sharply circumscribed. Pneumoencephalography in tumours of the gasserian ganglion reveals an elevation of the temporal horn and, in larger tumours, a displacement and shift of the posterior fossa structures.

Although Jefferson (1955) felt that cerebral arteriography was not diagnostic in these tumours, others since then have stressed the value of carotid and vertebral arteriography (Hauge, 1954; Holman *et al*, 1961; Ruberti, Galligioni, and Carteri, 1966). The characteristic features seen in the carotid arteriogram are a displacement of the ganglial part of the carotid siphon medially, forward, and

TABLE I
TRIGEMINAL NEURINOMAS IN ADOLESCENTS

Author	Sex and age (yr)	Duration of symptoms (yr)	Facial pain	Subjective sensory change in face	Objective findings of 5th nerve		Location of tumour	Posterior fossa symptoms	Surgical approach	Recurrence (yr)
					Sensory	Motor				
Shutova	F 18	2	0	Tingling	1, 2, 3 +++	++	?	+	Temporal	
Jefferson (case 3)	F 18	3	Early +	Paraesthesia and numbness (3)	2, 3 ++	0	3rd trigeminal		Temporal	
Jefferson (case 7)	M 20		+	Numbness	++	0	Posterior fossa (root)	+	Suboccipital	20
Olive and Svien (case 2)	M 21	4	0	Numbness (2)	1, 2, 3 +++	+++	Gasserian ganglion	—	Temporal	2
Olive and Svien (case 13)	M 21	4	0	'Stiffness' of face	1 +	0	5th nerve root	+	Suboccipital	
Schisano and Olivecrona (case 6)	F 22	11	0	?	++	?	Gasserian ganglion	+	Temporal	
Schisano and Olivecrona (case 9)	F 14	2½	0	?	1 +	?	Gasserian ganglion		Temporal	8
Gordy	F 17	1	0	Numbness	3 +++	+++	Gasserian ganglion		Temporal	
Case 1	M 14	2	Retro-orbital	Numbness (1, 2)	1, 2 +++	+++	Gasserian ganglion	+	Temporal	2
Case 2	M 16	2	0	Numbness (1, 2, 3)	1, 2, 3 +++	+++	Gasserian ganglion	+	Temporal	
Case 3	F 13	5	0	0	1 +	+++	Gasserian ganglion	0	Temporal	

downward and elevation of the superior portion of the siphon. The anterior choroidal artery, the Sylvian vessels, and the basal vein are also elevated. Vessels originating from the extradural portion of the carotid artery and entering the tumour were recognized and found enlarged by Westberg (1963). A 'feathery' tumour stain in the later phase was described (Gordy, 1965). In the vertebral arteriogram, the basilar artery is displaced posteriorly and medially and the posterior cerebral and superior cerebellar arteries are elevated and bowed, depending on the size and extension of the tumour. Khilnani and Silverstein (1963) studied the superior cerebellar arteries and found a medial and upward displacement (AP view) of the first portion of that artery only in the presence of a cerebello-pontine angle mass and considered this diagnostic for an extra-axial mass lesion. Some features described above are not specific for trigeminal neurinoma alone but occur with other tumours in this location in the middle fossa and in the cerebello-pontine angle. The cases reported showed bony erosion of the petrous bone and, on arteriography, the characteristic findings of displacement of major vessels, abnormal vessels, and tumour stain (Figs. 1 to 5).

Recently, Gass (1969) observed an early fifth nerve involvement by a tumour in spite of negative contrast studies and introduced the pantopaque basal cisternogram as another diagnostic test. This shows the filling defect of the normal trigeminal nerve root and also small tumours in the posterior fossa or cerebello-pontine angle. The demonstration of small lesions is important for early removal of the tumour and may be imperative in deciding the surgical approach.

Both cases represented large tumours with considerable extension into the posterior fossa. Nevertheless, a temporal craniotomy for approach to the tumour was chosen. The temporal lobe was elevated after opening the dura and the tumour was removed piecemeal intracapsularly. The tentorium was then sectioned, allowing easy access to the extension in the posterior fossa. This approach proved very effective and was stressed in previous reports (Jefferson, 1955; Loew and Tönnis, 1954; Olive and Svien, 1957; Schisano and Olivecrona, 1960). Complete removal of the tumour should be attempted to prevent a recurrence (case 1; Table I), although in some instances this will not be feasible and could result in irreparable damage to the internal carotid artery, the cavernous sinus, or the brain-stem. The use of the operating microscope should greatly facilitate removal of this tumour.

SUMMARY

Two cases of trigeminal neurinoma in adolescents

are reported. The pertinent literature revealed the significant incidence of 10% of these tumours occurring in adolescence. The characteristic symptoms are paresis of the fifth nerve (sensory and motor function) and involvement of other cranial nerves, the cerebellum, and the cerebral peduncles with the growth of tumour into the posterior fossa. Destructive changes in the floor of the middle fossa and erosion of the petrous apex are the abnormal features frequently seen in skull radiographs. We like to stress the value of carotid and vertebral arteriography revealing the typical displacement of the carotid siphon and major cerebral vessels and a tumour stain. The advantages of a temporal craniotomy are pointed out. Section of the tentorium may facilitate complete removal of large hourglass-shaped tumours by this approach.

ADDENDUM

After submitting the paper for publication, another trigeminal neurinoma in an adolescent was encountered (P.N.T.).

A 13-year-old girl was admitted with complaints of diminution of vision, intermittent redness of the left eye, and progressive thinning of the left face for the past five years. Abnormal findings on examination were limited to the left side with a decrease of visual acuity to finger counting, a dense corneal opacity, and absent corneal sensation. There was also a partial third, fourth, and sixth nerve paresis and complete motor fifth nerve palsy with marked wasting of the masticatory muscles. Sensation of the rest of the face was normal and no other motor, sensory, or cerebellar signs were present.

Skull films revealed a grossly enlarged sella and destruction of the apex of the left petrous bone. A left carotid arteriogram showed a large mass in the middle cranial fossa and pneumoencephalography outlined the extension of this tumour into the left cerebellar pontine angle. Cerebrospinal fluid protein was 220 mg%. The tumour was removed through a left temporal craniotomy and the pathological diagnosis of trigeminal neurinoma confirmed.

REFERENCES

- Altmann, F. (1928). Zur Kenntnis der primären Geschwülste des Trigemini und des Ganglion Gasseri. *Beitr. path. Anat.*, **80**, 361-404.
- Best, P. V. (1968). Erosion of the petrous temporal bone by neurilemmoma. *J. Neurosurg.*, **28**, 445-451.
- Gass, H. (1969). Unilateral numbness of the face and the basal cisternogram. *Neurology (Minneapolis)*, **19**, 66-70.
- Glaser, M. A. (1935). Tumors arising from the sensory root of the trigeminal nerve in the posterior fossa. Perineurial fibroblastoma. *Ann. Surg.*, **101**, 146-155.
- Gordy, P. D. (1965). Neurinoma of the gasserian ganglion. Report of a case and review of the literature. *J. Neurosurg.*, **22**, 90-94.
- Hauge, T. (1954). *Catheter Vertebral Angiography*, pp. 166-168. G. Lindkvist-Boktrykheri: Oslo.

- Holman, C. B., Olive, I., and Svien, H. J. (1961). Roentgenologic features of neurofibromas involving the gasserian ganglion. *Amer. J. Roentgenol.*, 86, 148-153.
- Jefferson, G. (1955). The trigeminal neurinomas with some remarks on malignant invasion of the gasserian ganglion. *Clin. Neurosurg.*, 1, 11-54.
- Khilnani, M., and Silverstein, A. (1963). Displacement of the superior cerebellar artery. *Arch. Neurol. (Chic.)*, 8, 502-505.
- Krayenbühl, H. (1936). Primary tumors of the root of the fifth cranial nerve: their distinction from tumors of the gasserian ganglion. *Brain*, 59, 337-352.
- Loew, F., and Tönnis, W. (1954). Klinik und Behandlung der Neurinome des Nervus trigeminus. *Zbl. Neurochir.*, 14, 32-41.
- Nowotny, K., and Uiberall, H. (1934). Zur Kenntnis der Neurinome des Trigemini. *Z. ges. Neurol. Psychiat.*, 150, 75-99.
- Olive, I., and Svien, H. J. (1957). Neurofibromas of the fifth cranial nerve. *J. Neurosurg.*, 14, 484-505.
- Ruberti, R., Galligioni, F., and Carteri, A. (1966). Considerations radiologiques dans les neurinomes du ganglion de Gasser. *Acta radiol. Diagn.*, 5, 465-469.
- Russell, D. S., and Rubinstein, L. J. (1963). *Pathology of Tumours of the Nervous System*, pp. 242-257. Arnold: London.
- Schisano, G., and Olivecrona, H. (1960). Neurinomas of the gasserian ganglion and trigeminal root. *J. Neurosurg.*, 17, 306-322.
- Shutova, T. A. (1940). Case of neurinoma originating in root of trigeminal nerve. *Nevropat. i Psichiat.*, 9, 109-111.
- Taveras, J. M., and Wood, E. H. (1964). *Diagnostic Neuroradiology*. Williams and Wilkins: Baltimore, Md.
- Westberg, G. (1963). Angiographic changes in neurinoma of the trigeminal nerve. *Acta radiol. Diagn.*, 1, 513-520.
- Zülch, K. J. (1965). *Brain Tumors. Their Biology and Pathology*. Springer: New York.