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Neurinoma of the Gasserian Ganglion and the Trigeminal Root

Report of Four Cases

By

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With 1 Figure

Summary

Four cases of neurinoma of the Gasserian ganglion and the trigeminal root are reported. The symptoms are analysed and compared with cases from the literature. The value of early operation in cases with a typical clinical course is emphasized, and it is recommended that operation is performed even before the lesion can be demonstrated by radiography.

Neurinomata of the trigeminal ganglion and root are relatively rare. In 1964 Krohm and Marguth traced 79 cases reported in the literature. They added 13 cases of their own, which represented 0.33 per cent of 4000 intracranial tumours. Roughly the same incidence was reported by Schisano and Olivecrona (1960), who observed 15 cases among 5727 brain tumours (0.26 per cent).

The purpose of this paper is to analyse four cases of trigeminal neurinoma seen in our department in the five-year period from April 1965 to April 1970.

Patients

Case 1. A 36-year-old man was first seen in September 1965. He complained of numbness of the right side of the face which had persisted for 11 months. For 5 months he had attacks of pain in the occipital region provoked by changes in posture, coughing and sneezing, and for 6 weeks episodes of projectile vomiting and dizziness. Diplopia had been present for the last 3 weeks.

Examination. He was clear and co-operative, but suffered from a severe psychoneurosis with anxiety and depression. Right oculomotor palsy and

hypo-aesthesia of the right trigeminal area were observed, whereas the remaining cranial nerves were intact, and there were no cerebellar or pyramidal-tract signs. There was no paralysis of the masticatory muscles. The optic discs were normal. Pure-tone audiogram, caloric tests, electro-encephalogram, radiographs of the skull and temporal bones, right carotid angiogram and pneumo-encephalogram were all normal. The protein content of the lumbar cerebrospinal fluid was 85 mg/100 ml; pressure 260 mm of fluid. During the next 4 months, symptoms and signs of cerebellar and pyramidal-tract involvement developed, accompanied by increasing drowsiness. *Right carotid angiography* and *ventriculography* showed an hour-glass tumour with the larger portion in the posterior fossa.

Operation. In February 1966, suboccipital craniotomy was performed with a wide right cerebellar exposure. On retracting the right cerebellar lobe a large tumour was seen. The trigeminal nerve was not in view, but the 7th and 8th nerves shirted the lower pole of the tumour, which had the appearance of a neurinoma. As was anticipated from the angiogram the tumour had displaced the carotid artery. The 3rd and 4th nerves were embedded in the thick capsule and were both damaged when the capsule was removed.

Pathology. Histological examination of the tumour revealed all the features of a typical cystic neurinoma. In a few areas, myelinated nerve fibres were seen, a couple of them being infiltrated by the tumour.

Postoperative course. The operation was followed by palsy of the right 3rd and 4th nerves, paralysis of the right masticatory muscles, complete anaesthesia in the area of the ophthalmic and maxillary divisions of the right 5th nerve, total deafness and absence of caloric reactions of the right ear. A right peripheral facial palsy recovered during the first few months.

At the most recent follow-up examination 6 years after the operation the patient was working full-time as an engineer.

Case 2. A 53-year-old man was admitted to our department in January 1966, complaining of numbness in the forehead, cheek and upper lip on the right side of the face, which had persisted for 14 months. Occasionally, he had experienced paroxysms of atypical facial pain in the same areas. For a period of 2 months he had been suffering from constant vertigo, tinnitus of the right ear, diplopia, difficulty in swallowing because of regurgitation into the nose and laryngeal spill-over, paresis of the left upper and lower limbs and increasing loss of recent memory.

Examination. He was apathetic with delayed responses. There were incomplete palsy of the right abducens nerve, hypo-aesthesia of all three divisions of the right 5th nerve, and paresis of the right muscles of mastication and of the soft palate, but no facial palsy. The optic discs were normal. There was a left-sided slightly spastic hemiplegia. Pure-tone audiogram showed slight symmetrical high-tone deafness with abnormal tone decay in both ears. There was spontaneous nystagmus to the left, which could be changed by caloric stimulation, but 10 days later complete deafness in the right ear developed, and a vestibular response could no longer be elicited.

Carotid angiography, pneumo-encephalography, electro-encephalography, radiography of the skull and the temporal bones were all normal. The protein content of the cerebrospinal fluid was 70 mg/100 ml.

Operation. In January 1966, suboccipital craniotomy was performed with a wide right cerebellar exposure. A cystic neurinoma was present in the right

cerebellopontine angle. The tumour was very soft; the 7th and 8th cranial nerves were lightly embedded in it, but could easily be separated. The tumour extended down between pons and clivus and evidently arose from the root of the right trigeminal nerve.

Pathology. Histological studies revealed a neurinoma. Demyelinated nerve fibres were seen in close contact with the tumour.

Postoperative course. A few days after the operation the patient was alert. The above-mentioned cranial nerve defects persisted, whereas the weakness of the limbs gradually disappeared. At a follow-up examination 5 years after the operation the patient had no facial pain, vertigo or tinnitus, but he had been unable to resume his usual work as a bricklayer because of persistent loss of memory.

Case 3. A 16-year-old girl was admitted to our department in December 1966, complaining of headache and tinnitus, which had been present for 3 weeks and of nausea and vomiting of a few days duration.

Examination. The general physical and neurological examination did not disclose any abnormalities, but ophthalmoscopy revealed choked discs. Electro-encephalography showed focal abnormities in the left temporal region.

Left carotid angiography, left vertebral angiography and *ventriculography* showed a subtemporal tumour.

The protein content of the cerebrospinal fluid was 54 mg/100 ml., pressure 800 mm of fluid.

Operation. In December 1966, the left temporal lobe was elevated through a flap incision. This revealed a very thin-walled multicystic tumour containing clear, yellow fluid. When the cysts had been emptied and removed, a solid tumour was seen. It extended into the posterior fossa in front of the pons and was intimately connected to the root of the trigeminal nerve. The removal of this portion of the tumour was attended with some difficulty.

Pathology. Histological examination revealed a neurinoma. Non-myelinated nerve fibres were seen in one area.

Postoperative course. This was satisfactory during the first week. The patient was alert with hypo-aesthesia in the first division of the left trigeminal nerve and a right abducens paralysis. However, she became increasingly drowsy during the next few days, and left carotid angiography now aroused suspicion of a subdural haemorrhage. Re-operation performed 10 days after the first operation showed only cerebral oedema and a subdural hygroma, but no haemorrhage. Subsequent check-up carotid angiography revealed severe arterial spasm, but did not support the suspicion of tumour or haemorrhage. The patient remained comatose until she died of pulmonary complications 2 months after the operation.

Autopsy revealed diffuse cortical necrosis. No residual tumour tissue was seen.

Case 4. A 31-year-old woman was admitted to our department in December 1969. For 5 years she had suffered from headache—in the first 3 years migraine-like with periodic attacks of left frontal headache associated with nausea and photophobia, in the last 2 years constant pain in the left temporal region. During the last 12 months she had experienced diplopia and numbness of left frontal region and cheek, and deviation of the jaw had been present.

Examination. The only positive signs were impairment of the left 5th and 6th cranial nerves. Hypo-aesthesia in the areas supplied by the ophthalmic

and maxillary divisions of the left trigeminal nerve, paresis of the masticatory muscles, and paralysis of the left abducens nerve. The optic discs were normal. Pure-tone audiometry, caloric test, radiography of the skull, and tomography of the temporal bones were normal. Electro-encephalography was slightly abnormal with theta frequencies in both temporal-occipital regions and a slight left-sided preponderance. The protein content of the lumbar cerebrospinal fluid was 40 mg/100 ml. Left carotid angiography and ventriculography showed normal conditions.

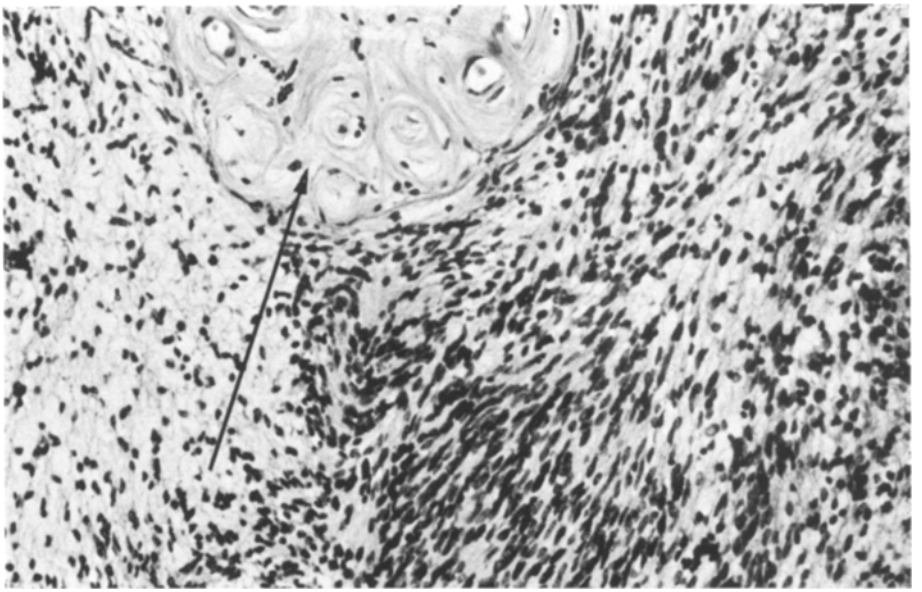

Fig. 1. Case 4, neurinoma with spindle cells and foam cells. Arrow indicates blood vessels with typical tickened collagenous hyalinized walls. Haematoxylin and eosin, $\times 160$

Operation. In December 1969, the left temporal lobe was elevated through a temporal flap incision. This revealed a small tumour beneath the dura at a depth of 3 cm. Incision of the dura showed a soft cystic tumour centred in the ganglion of the 5th nerve. The nerve fibres of the divisions were stretched and displaced by the tumour. There were no signs of extension into the posterior fossa.

Pathology. Typical cystic neurinoma. Ganglion cells with degeneration and myelinated nerve fibres were seen in a few areas of the specimen.

Postoperative course. The patient made a quick recovery; her headache and facial pain disappeared, but the cranial nerve defects persisted. At a follow-up examination 2 years after the operation she had still hypo-aesthesia in the maxillary division of the trigeminal nerve, but not in the ophthalmic division, and she had no diplopia. She was able to look after her house unaided.

Comments

The principal problem of trigeminal neurinoma is late diagnosis. The patients often have symptoms for many months or several years before the diagnosis is established, and at the time of operation the tumour has in many cases reached a size that makes radical surgery impossible.

Most of the cases reported in the literature show a characteristic clinical picture. The outstanding feature is that the initial symptoms are referable to the fifth cranial nerve, and later adjacent cranial nerves are affected.

Hypaesthesia of the face is more common than facial pain. Jefferson (1955) emphasized that complete anaesthesia is more suggestive of malignant Gasserian tumour than of a benign neurinoma. In cases with facial pain there are usually atypical non-paroxysmal neuralgias, only very rarely like the type of tic douloureux (Jefferson 1955, Rasmussen 1965). According to Krayenbühl (1936) and Schisano and Olivecrona (1960), constant facial pain is suggestive of a tumour of the ganglion, while root tumours only rarely cause pain. It is remarkable that only in 8 per cent of the cases reported in the literature was the 5th nerve intact at the time of operation (Krohm and Marguth 1964).

Tumours arising from the Gasserian ganglion may affect the 3rd, 6th and 4th cranial nerves resulting in diplopia. Tumours of the trigeminal root may affect the 3rd, 4th, 6th, 7th and 8th cranial nerves. In 10%—15% of the cases, impairment of hearing or vertigo is the first symptom; it is present on admission to hospital in about 30%, and in these cases differentiation from acoustic neurinoma may be impossible. Affection of the brain stem and cerebellar hemispheres by compression are late signs.

The most important features in radiographs of the skull are erosion of the foramen ovale, foramen rotundum and the petrous apex (Lindgren 1941), but these are late signs. In most cases from the literature, pneumo-encephalography and carotid angiography were performed; in some cases also vertebral angiography; but it is evident that the tumour must reach a certain size before it can be demonstrated by these methods.

In our small series, cases 1, 2, and 4 followed the common clinical course, whereas case 3 was rather unusual, both because of the early intracranial hypertension and the early age. Pneumo-encephalography, carotid and vertebral angiography were of value in cases 1 and 3, but as indicated by the clinical course, case 1 should have been subjected to operation shortly after the first admission, just like cases 2 and 4. The radiographs of the skull were normal in all four cases. It is remarkable that the protein content of the cerebrospinal fluid was normal or only slightly increased in our cases.

We are convinced that all patients with a typical clinical course should be subjected to operation on clinical evidence when the tumour is small, before erosions are visible in the radiographs, and before the tumour becomes so large that it can be demonstrated by pneumoencephalography or angiography.

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