Erosion of the Petrous Temporal Bone by Neurilemmoma*

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Apart from the well-recognized widening of the internal auditory meatus by neurilemmoma of the 8th cranial nerve, this type of tumor rarely causes significant erosion of the temporal bone. Such behavior is, however, one of the main features of the very uncommon 5th nerve neurilemmomas (Olive and Svien,8 Schisano and Olivecrona10). In order that adequate treatment may be undertaken it is important to recognize that these benign tumors can erode or even infiltrate bone.

This report presents three cases of petrous erosion by neurilemmoma. The first case was that of a combined supratentorial and infratentorial neurilemmoma probably arising from the trigeminal nerve (Jefferson9), the second was a neurilemmoma almost entirely confined to the petrous bone, and the third* was exceptional in that the intratentorial growth was combined with a mass in the cerebellopontine angle and infiltration into the middle-ear and external auditory meatus.

Case Reports

Case 1. A 59-year-old man developed hallucinations and cerebellar signs while hospitalized for acute pulmonary manifestations of long standing tuberculosis. He had been employed as a stone mason and developed pulmonary tuberculosis in 1952, possibly associated with silicosis. He had been admitted to hospital several times for treatment, which included thoracoplasty in 1956. In spite of this the disease had gradually progressed until the patient became a respiratory cripple. His final admission to hospital was on July 6, 1965, again due to increasing dyspnea and productive cough with hemoptysis. He then began to develop a peculiar mental state with hallucinations and delusions of persecution, claiming that other patients were "ganging up on him." At times he became confused and aggressive, but all these symptoms subsided on treatment with tranquilizing drugs. On October 15, he complained of having seen "flashing lights" during the previous night.

Examination. The patient had a very ataxic gait and coarse nystagmus on right lateral gaze. Ataxia was also shown on finger-nose and heel-knee testing, slightly more marked on the right than the left. The right corneal reflex may have been slightly diminished, and there was slight nerve deafness on the right. The protein content of the lumbar cerebrospinal fluid was 550 mg/100 ml. Tomographic x-rays of the skull showed a normal left internal auditory meatus, but on the right there was loss of the petrous tip and destruction of the internal auditory canal (Fig. 1). A diagnosis of right cerebellopontine angle tumor was made, but the patient’s general condition was too poor to allow surgical treatment.

The neurological symptoms and signs improved during the next 2 weeks by which time the ataxia had completely disappeared. The patient still had slight deafness with tinnitus on the right, slight nystagmus to the right, and equivocally diminished right corneal reflex, but no other abnormalities were detected. Cough and hemoptysis recurred on December 6, with increasing difficulty in breathing. There was clinical evidence of bronchopneumonia. In spite of treatment the

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FIG. 1. Case 1. X-ray showing erosion of right petrous tip.
Case 2. A 32-year-old woman was admitted to the Midland Centre for Neurosurgery and Neurology, Smethwick, Staffordshire, with a history of increasing deafness in the right ear for 10 years and total deafness for about 1 year. For 4 months she had had morning headaches, unaccompanied by vomiting, and attacks of numbness and “stiffening” of the right side of the mouth lasting 2 to 3 minutes about three times a day. In the last 4 weeks there had been dizziness and blurring of vision of the right eye, especially on movement.

Examination. The patient had gross bilateral papilledema with hemorrhages, worse on the right than the left, nystagmus in all directions but most coarse to the right, a diminished right corneal reflex, impaired sensation over the area supplied by the first and second divisions of the right 5th nerve, complete right nerve deafness, and a bruit over the right mastoid when the head was in full flexion. General examination revealed no significant abnormalities. A full blood count and chest x-rays were normal. Audiogram and caloric tests showed complete loss of function on the right. Skull x-rays showed extensive erosion of the right petrous apex (Fig. 3), and a right carotid arteriogram showed forward displacement of the internal carotid artery at this level.

Operation. Craniotomy was carried out through a right suboccipital approach (Mr. J. M. Small). In spite of previous ventricular tapping, the dura was tense. The right cerebellar tonsil was herniated, but some fluid was obtained from below it. Retraction of the cerebellum exposed an extradural tumor bulging into the cerebellopontine angle so
that the 7th and 8th nerves could not be seen. After the dura was incised, a firm, fleshy, partly calcified tumor was entered, the appearances of which suggested a chondroma with probable sarcomatous change. A large amount of it was removed by working extradurally within the petrous bone. The dura was left open and the incision closed in layers without drainage. Histologically the tumor proved to be a typical benign neurilemmoma (Fig. 4).

The patient made a satisfactory postoperative recovery, but the papilledema remained and the skin over the posterior fossa was tense.

Second operation. Since the tumor had been shown to be benign, the area was reexplored by Mr. Small 5 weeks after the first operation. The muscle flap was adherent to the surface of the cerebellum but sufficient exposure was obtained to allow removal of the lateral third of the cerebellar hemisphere and give access to the tumor. The residual cavity in it was reentered and a larger removal of neoplastic tissue accomplished; the majority of the tumor tissue was well forward in the petrous bone, evidently very close to the carotid artery. Dura which pulsated inwards could be seen in the roof and posteromedial wall of the new cavity. The anterosuperior surface of the cerebellum was adherent to the dura and capsule; it was considered inadvisable to go further medially towards the pons extradurally because of the risk of encountering uncontrollable hemorrhage. The whole tumor was apparently extradural within the petrous bone and at no time were the 5th, 7th, or 8th nerves seen.

Postoperative course. The patient had only slight sensory loss and weakness of the face; the main abnormalities were ataxia on the right and impaired vision probably due to the previous papilledema. Ventriculography was carried out to ensure that there was no further obstruction, and the patient was making a satisfactory recovery when she was discharged from the hospital 11 weeks after admission.

Case 3. A 46-year-old woman was first admitted to the Ear, Nose, and Throat Department, Aberdeen Royal Infirmary, on May 31, 1959 (under the care of Mr. C. D. Weir) with a history of deafness and “buzzing” in the left ear for about 10 years and discomfort in the ear for 1 month.

A firm polyloid mass occluding the left external auditory meatus but with no attachment to the bone walls was incompletely removed on June 1, 1959; it was then noted that the polyp had grown from the promontory through a large perforation in the tympanum. Histological examination showed a spindle-cell tumor which was diagnosed as a sclerosing hemangioma. The patient was kept under regular observation as an outpatient for 18 months, after which she defaulted in her attendance. There had, however, been no evidence of recurrence up to that time.

The patient was next seen by the Ophthalmology and Neurosurgical Departments in May, 1963, because of a 6-month history of attacks of occipital headache with radiation over the vertex to the left eye. The pain was accompanied by vertigo and blurring of vision in the left eye, and there had been some general deterioration of visual acuity.

Examination. There was bilateral papilledema, left facial weakness, and left-sided deafness. There were no sensory changes in the face and no cerebellar signs. The polyloid mass in the left ear had reappeared and grown half way along the external meatus. The only abnormality found on general examination was that the blood pressure was
210/120 mm Hg whereas it had been normal in 1959. Audiometry showed conduction deafness and moderate nerve deafness. Tomographic skull x-rays (Dr. R. A. McKail) revealed a sharply defined area of erosion in the left petrous bone (Fig. 5) involving the internal auditory meatus and extending downward and sideways. At one point the bone destruction appeared to extend through to the base in the region of the jugular fossa. The cochlea and semicircular canals could not be identified.

Operation. Myodil ventriculography on June 4, 1963, revealed deviation of the aqueduct and body of the fourth ventricle to the right; exploration of the posterior fossa was carried out the same day (Mr. W. Martin Nichols). The right lateral ventricle was first cannulated to permit decompression. In the left cerebellopontine angle a tumor was found that had the appearance of an acoustic neurilemmoma although it seemed to arise at a slightly lower site than usual. The capsule was incised and much of the tumor removed piecemeal. During this procedure the capsule tended to collapse medially as if the tumor had arisen from the floor of the posterior fossa rather than from the lateral wall. Complete extirpation was not attempted because of the unknown nature of the tumor and the known extent of infiltrative growth. There was considerable bleeding but this seemed to have been adequately controlled before closure.

The patient's immediate postoperative progress was satisfactory; she regained consciousness, spoke, and moved all her limbs. Later the same day, however, sudden respiratory arrest occurred followed by cardiac arrest, and all attempts at resuscitation failed.

Necropsy. Apart from the intracranial abnormalities, the only pathological finding of note was a small cavitated lesion in the lower lobe of the left lung, which proved on subsequent histological examination to be a squamous carcinoma. There was no evidence of spread to lymph nodes or other tissues either macroscopically or histologically. A tumor measuring \(3.5 \times 2.5 \times 2.5\) cm was present in the left cerebellopontine angle (Fig. 6); it had a smooth gray capsule with yellowish patches. Transection after fixation revealed that most of the material within the capsule was blood clot and there was very little residual neoplastic tissue. A prominent foraminal pressure cone was present, and there was mild symmetrical hydrocephalus.

When the dura was stripped from the base of the skull, small grayish-white, dome-shaped tumor nodules were seen projecting through the tegmen tympani (Fig. 7). A wedge of bone was cut from this region of the base of the skull, decalcified, and transected. This showed that the tumor filled the middle ear cavity, mastoid air cells, and medial part of the external auditory meatus, extending downward to the region of the jugular fossa (Fig. 8). Definite continuity could not be seen between the intrapetrous tumor and the cerebellopontine angle tumor, but there was considerable hematoma in the vicinity of the
internal auditory meatus where a large portion of the tumor had been surgically removed.

Histologically all parts of the tumor showed the typical features of a benign neurilemmoma (Fig. 9). Review of the sections of the biopsy specimen taken from the external auditory meatus 4 years earlier revealed an identical neoplasm had been present beneath the subepithelial connective tissue (Fig. 10). There was infiltration of bone by the tumor. The intra-osseous and meatal part of the neoplasm was formed predominantly of the compact, Antoni type A cellular structure, whereas in the cerebellopontine angle tumor the looser textured type B architecture was also a prominent component.

Discussion

Petrous erosion is one of the most important diagnostic features of those very uncommon neurilemmomas that arise from the trigeminal nerve (Olive and Svien, Schisano and Olivecrona). The site of the tumor in Case 1 corresponded closely with that described for the combined supratentorial and infratentorial forms of trigeminal neurilemma. The exact site of origin, however, could not be identified and the clinical presentation was very unusual. The first neurological disturbance appears to have been mental change, followed by transient ataxia. The only other abnormalities were mild ipsilateral deafness with tinnitus, slight nystagmus, and equivocal depression of the corneal reflex. In spite of the considerable size of the mass, it had only produced these relatively minor clinical effects in the last 5 months of the patient's life, death being due to preexisting respiratory disease. The sole clinical indication of 5th nerve involvement was the doubtful depression of the corneal reflex. That this does not exclude the possibility of a trigeminal origin has been emphasized in the reports cited above.

The assessment of the probable nerve of origin in the Case 2 tumor is more difficult. Here the mass was extradural and entirely within the petrous bone except for the bulge...
into the cerebellopontine angle which had produced intracranial pressure effects. A similar intrapetrous growth in a 16-year-old girl who had had deafness for 8 years and facial weakness for 2½ years was reported by Love in 1950; he considered that the tumor probably had arisen from the 8th nerve. In 1966 Binns and Fairman described another similar example (Case 3) in a young woman who had clear evidence of trigeminal involvement with deafness as a minor feature. The long history of increasing deafness and late onset of sensory symptoms in the face suggest that the tumor in our Case 2 arose from the 8th nerve rather than the 5th. An interesting possibility is that it may have originated in the cochlea; Jørgensen reported a case of intracochlear neurinoma in 1962.

The behavior of the tumor in Case 3 was extremely unusual, the initial presentation being the growth within the external auditory meatus. A strikingly similar pattern of neoplasia described in 1959 by Pou (his Case 2) was attributed to separate origins from the 7th and 8th nerves. In Case 5 of Olive and Svien, a predominantly intrapetrous neurilemmoma also extended into the external auditory canal; although there was no cerebellopontine angle tumor, this patient had had ipsilateral deafness for 4 years and evidence of 5th nerve involvement for 8 months. The authors included the case in their group of supratentorial trigeminal tumors, but the evidence given for the site of origin seems inconclusive. Facial nerve neurilemmomas, which are even less common than the trigeminal ones, are particularly liable to extend into the middle ear and external auditory meatus (Altman and Furlow). Facial weakness is usually the first sign; this was a very late development in our Case 3. An 8th nerve or perhaps intracochlear origin for the tumor in our Case 3 seems more likely; this may also have been the origin of the tumor in Pou’s Case 2 and Olive and Svien’s Case 5.

It is interesting that, excluding the typical neurilemmomas of trigeminal origin, tumors of this nature associated with intrapetrous growth seem to occur predominantly in young women.

From the pathological point of view the behavior of some of these intrapetrous tumors is of considerable interest. Although they apparently are all of histologically typical, benign neurilemmomatous structure, in rare instances these tumors show a potentiality for infiltration of bone. This behavior is similar to that of some meningiomas, raising the possibility of an infiltrative growth that cannot be predicted on histological grounds alone. The great majority remain as encapsulated growths; if bone erosion occurs it is presumably due to pressure atrophy, but there are these rare instances of invasive growth by tumors indistinguishable histologically. It is important to realize that these intrapetrous tumors may well be amenable to surgical treatment, as illustrated by Case 2, even though complete excision is likely to be difficult if not impossible.

Summary

Three cases in which erosion of the petrous temporal bone was caused by a neurilemmoma have been presented. In one, the features were
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typical of a trigeminal origin, with part of the mass below and part above the tentorium. In another, the growth was confined to the petrous bone apart from the bulge it produced into the cerebellopontine angle. In the last case there was intra-osseous growth in addition to a mass in the cerebellopontine angle and an extension into the external auditory meatus. In these last two cases an origin from the 8th nerve or cochlea seemed likely.

If typical trigeminal neurilemmomas are excluded, reported cases of intrapetrous tumors of this nature have occurred mainly in young women.

The neurilemmoma that infiltrates bone may have a benign histological appearance indistinguishable from that of the more usual, encapsulated form.

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References