Intracranial hypoglossal neurinomas—
A report of two cases

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INTRACRANIAL NEURINOMAS of the hypoglossal nerve are indeed rare in the experience of most neurosurgeons. A review of the literature by Ignelsi and Bucy in 1967 revealed only nine documented cases of this tumor of the hypoglossal nerve. They added a tenth case to the literature. At that time, these authors emphasised the need for elective tracheostomy following surgical extirpation of the tumor, if the high surgical mortality and morbidity were to be reduced. Their patient survived in spite of being seen late in the course of his disease.

We would like to report two cases of hypoglossal neurinoma that were successfully treated by surgical extirpation of the tumor and elective immediate post-operative tracheostomy. Further, we would like to emphasise two points. Firstly, that during myelography the pantopaque column should be carried well into the cisterna magna if these tumors are to be recognised. If one were to be satisfied with a normal cervical myelogram, one would certainly miss diagnosing these tumors at myelography. Secondly, the possibility of the development of hydrocephalus post-operatively should be borne in mind. Delay in recognising this almost led to the demise of one of our patients.

Case Report I—J.S. NS: 5506/70

The patient, a 40-year-old Punjabi, was admitted into the medical service of this hospital on 17.2.70. He was referred to the Department of Neurosurgery on 22.2.70 when he gave the following history.

He had apparently been well until a year ago when he developed left-sided headaches, usually frontal and occipital in location, associated with nausea and a sense of vertigo whenever he climbed stairs. His work entailed using a ladder periodically and whenever he did so, he became unsteady on his feet and had a sense of spinning around. Two months prior to his admission, he noticed an unsteadiness of his gait associated with buzzing in his ears. At about this time, his speech also began to slur and he was troubled with pricking and pulling sensations in both his hands, especially at night. He had difficulty enunciating words and this had progressed to the point that he had great difficulty in making himself understood. Further he had begun to drool saliva at about this time.

His family and past history were unremarkable.

On examination, he was a well-built man with

Fig. 1: View of the tongues of patients in Case Reports I and II.

a pulse rate of 84 per minute, respirations 18 per minute, and blood pressure of 100/80 mm. Hg. There were no cafe au lait spots to be seen. His main findings were on examination of the central nervous system. Review of his various other systems revealed no abnormality.

Examination of the central nervous system revealed a conscious, rational and obviously dysarthric individual. His pupils were equal and reactive to light and accommodation. Funduscopy showed early bilateral papilledema. There was a horizontal nystagmus on looking to the left with the fast component to the left and mild weakness of his left 6th nerve. The most remarkable finding was a hemiatrophy of his tongue on the left side with marked fasciculations bilaterally. There was also flattening of the soft palate on the left but the gag reflex was intact. In addition to an intention tremor, he had a poor heel-to-knee test on his left side. Further there was considerable decomposition of movement on the left side. His gait was broad based and he was markedly ataxic with a tendency to fall to the left. Rhomberg sign was positive. His motor power was, however, good on both sides. His reflexes were brisk bilaterally but they were somewhat more so on the left side. There was no Babinski response. In addition, there was some increase in tone, especially in both lower limbs.

Pertinent investigations

Lumbar Puncture revealed an opening pressure of 210 mm. of C.S.F. The C.S.F. was clear and colorless.

Cythology revealed — 25 cells/cu. mm. mostly consisting of lymphocytes. The C.S.F. protein was 128 mg.%, globulin positive, sugar 62 mg.%, chlorides 120 meq/litre.

The following day, a pantopaque myelogram was undertaken which revealed a normal cervical myelogram. As we were suspecting a lesion of the 12th nerve posterior fossa, pantopaque cisternography was carried out, and this revealed a complete block to the dye column just rostral to the cisterna magna. The myelographic appearance was compatible with an intradural extramedullary mass lesion. Accordingly, a posterior fossa exploration was performed on 3.3.70.

A midline craniectomy was performed over the occipital bone following division and retraction of the cervical neck muscles. Laminectomy of C1 and C2 was also accomplished. Exploration of the

cerebello-medullary junction revealed a well circumscribed reddish yellow tumor measuring 1 x ¾ inches attached to the 12th nerve. About half of the tumor was disposed anterior to the medulla oblongata which had been pushed upwards and to the right by this tumor. The spinal accessory nerve was also stretched and pushed downwards and appeared to skirt the inferior pole of this tumor. An incision was then made over the tumor and its contents removed with forceps, spoons and suction. Following adequate decompression of the tumor contents, the capsule of the tumor was carefully delivered and dissected free from the medulla oblongata. It was then removed. Adequate care was given to preserve the blood supply to the medulla oblongata. Following hemostasis, the wound was closed in layers. The patient was then returned to the supine position and elective tracheostomy was performed. The patient was then returned to the intensive care unit.

Post-operatively, the patient had some difficulty in handling his secretion. For some four days, he drooled saliva and had to constantly resort to wiping his mouth dry. He otherwise made an uneventful recovery and by the fifth day post-operatively, he was able to handle his own secretions well and take oral fluids. A week later, he was able to walk with assistance and the tracheal stoma was closed. He was discharged from hospital on 24.3.70. At the time of his discharge, he continued to have some unsteadiness in his gait but he was ambulatory and well.

The patient was seen periodically at bi-weekly intervals from the time of his discharge. On his fourth visit, he complained of dizziness and a tight feeling in the back of his head and neck. Examination of the patient did not reveal any gross abnormality besides his unsteadiness which had been evident at the time of his discharge. Two days later, he was readmitted into the hospital acutely ill, complaining of severe headaches and pain in the nape of his neck and top of his head. In addition, he was nauseated and had had four episodes of projectile vomiting. When seen, he was irritable and violent. Examination now revealed early papilledema associated with a tense posterior fossa at the operational site. In addition, he was lethargic, resented examination and tended to curl up and sleep. However, when disturbed, he would react most boisterously and angrily. He was immediately taken to the operating room and after adequate preparation of his head and neck, a Right Ventriculo-Atrial shunt was planned. At operation, the intraventricular pressure was recorded at 360 mm. of

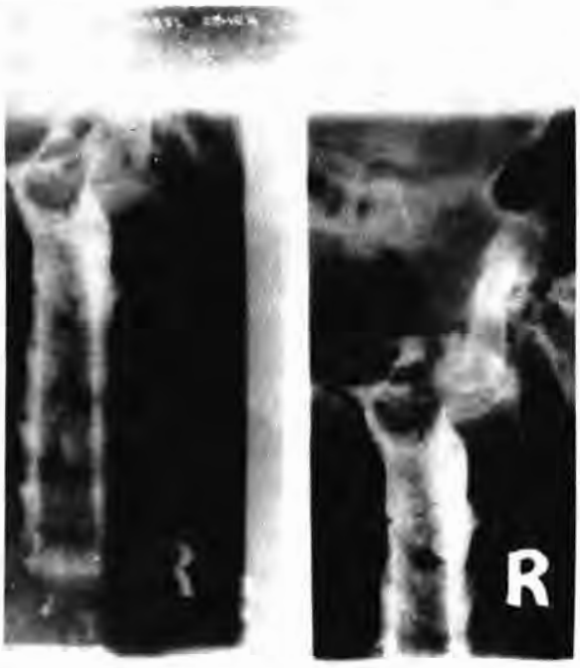

Fig. 2A: Normal cervical myelogram (Case Report I).

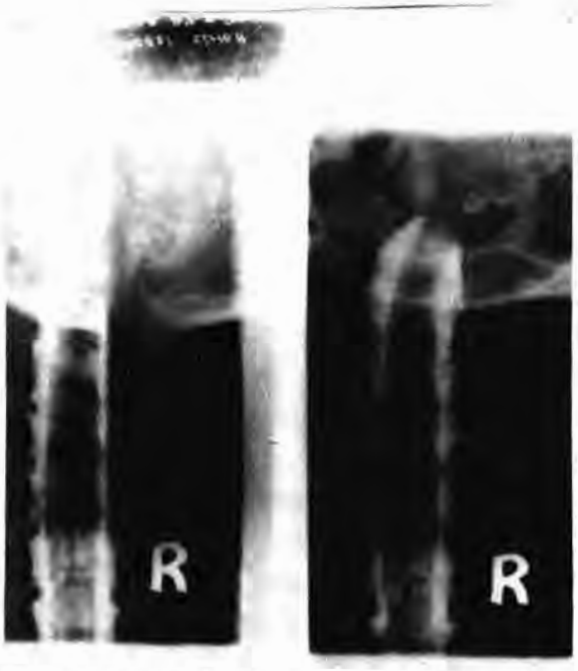

Fig. 2B: Dye in the cisterna magna now reveals the tumor.

cerebrospinal fluid. A right Ventriculo-Atrial shunt, with the aid of a Mischler Pudenz apparatus, was accomplished. Following the shunting procedure, the patient became quite calm and was his usual self again. He was discharged from hospital some eight days later in good condition. He is being followed on an outpatient basis at the present time.

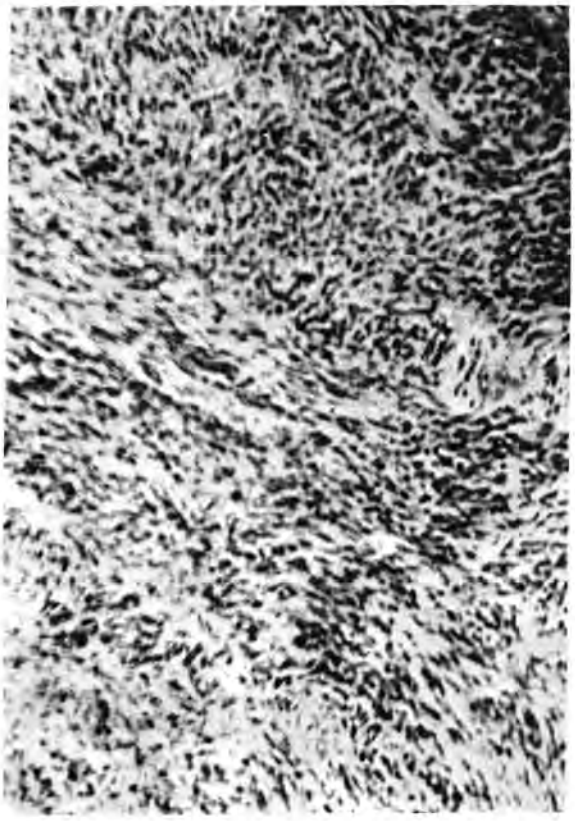

Fig. 3: Histology of the tumor (Case I).

Histopathology

Hematoxylin and Eosin preparations of the tumor removed at surgery revealed appearances fully compatible with those of a neurinoma.

Case Report II—T.P. NS: 6828/71

This 51-year-old Chinese woman was referred in from the Malacca General Hospital with a three-month history of headaches confined to the occipital area of her skull. In addition, she complained of numbness over the left side of her face and loss of taste on the right side of her tongue. Her symptoms began some four months previously and two weeks prior to admission to our center, she had begun to have difficulty in swallowing. On examination, she had a right 7th nerve palsy of the lower motor neuron type and marked wasting of the tongue on the right side. There was hyposthesia of the left side of the face and she had, in addition, corneal anaesthesia on this side. Her left soft palate did not move as well as on the right side. Reflexes were equal 1+, and there was no Babinski response. There were no cerebellar findings. A tentative diagnosis of 12th nerve neurinoma was made and a pantopaque posterior fossa study undertaken. A lumbar puncture to effect the study revealed an opening pressure of 180 mm. of water and a total protein of 114 mg.%, globulins were positive, sugar was 44 mg.%. The posterior fossa pantopaque study revealed evidence of tonsillar herniation with a complete high cervical block.

On 1.6.71, posterior fossa craniectomy was performed. The arch of the Atlas and the lamina of the 2nd cervical vertebrae were also removed. On opening the dura, the tonsillar herniation was confirmed and this was especially marked on the right side. On opening the dura over the cerebellar hemisphere, it was evident that there was a tumor in the region of the right cerebello medullary junction extending backwards between the right tonsil and the medulla. Tumor was greyish in color

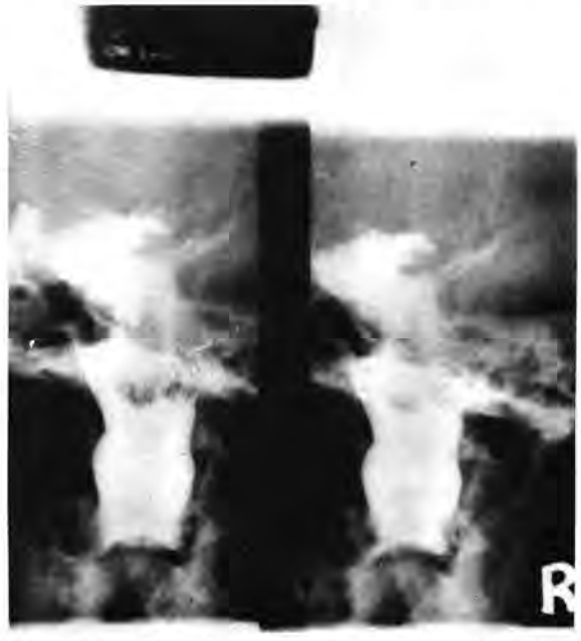

Fig. 4: Positivity of the pantopaque posterior fossa study is obvious (Case II). Note the filling defect secondary to the cerebellar tonsillar herniation.

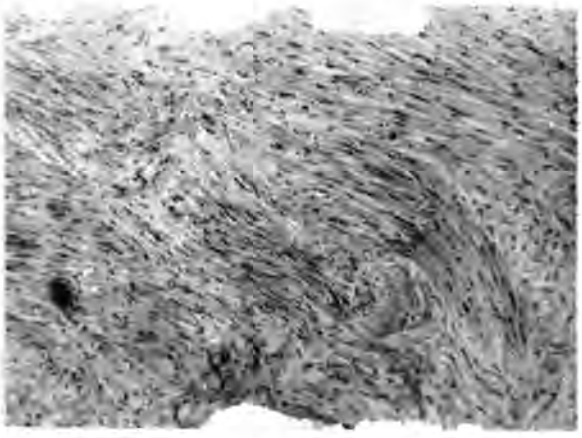

Fig. 5: Histopathology of the tumor (Case II).

and measured 1cm x 1½cm x 1½cm. It was well encapsulated and following internal decompression of the tumor, the posterior lateral and superior walls of the capsule were removed. The antero-medial wall attached to the medulla was left alone as it was considered not amenable to extirpation.

Post-operatively, the patient was subjected to elective tracheostomy for better respiratory management. At the time of her discharge from hospital three weeks later, she was ambulant and well. She was seen in follow-up a month later and she continues to do well.

Histopathology of the tumor removed showed appearances fully compatible with those of a neurinoma.

Summary

Two cases of intracranial neurinoma of the hypoglossal nerve successfully treated by surgical extirpation and elective immediate post-operative tracheostomy are reported. Careful high cervical myelography with the contrast material pushed well into the cisterna magna is necessary if these tumors are not to be missed. The possibility of acute hydrocephalus developing weeks to months post-operatively should be borne in mind if the condition is to be recognised early and treated promptly. The extreme rarity of these tumors in the world literature has prompted us to report these two cases.

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