Case Reports and Technical Notes

Spinocranial Neurofibroma

Case Report

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The occurrence of a neurofibroma at the level of the foramen magnum is rare. Of the 30 cases of foramen magnum tumors collected by Dodge, et al., only four, or 13%, were neurofibromas. Benign tumors in this area may attain considerable size before producing severe neurological deficits or a fatal outcome because of the relatively large subarachnoid space in the region of the upper cervical cord and lower medulla. Because of the proximity of these tumors to vital centers in the medulla, there is a considerable operative risk involved in their removal. In the following case, the entire tumor was successfully extirpated despite respiratory arrest during the operation. The patient recovered from the neurological deficit 8 months following operation.

Case Report

A 17-year-old girl was admitted to Soo Do Medical College Hospital on February 7, 1964, with the complaints of paralysis of all four limbs and difficulty with breathing. She had been well until 4 months earlier when she noted a tingling sensation first in the right arm, then shortly thereafter in the right leg. Within 2 months this became associated with motor weakness of the right arm and leg. Approximately 2 months before admission she noted a tingling sensation and paralysis in the left arm and leg. The quadriparesis was progressive and was associated with difficulty in voiding. She began to experience dyspnea 1½ months before admission which was also progressive in nature.

Examination. The pulse was 99, respiration 18, temperature 37.0, and blood pressure 120/60 mm Hg. There was flaccid paralysis of all extremities with loss of the superficial and deep reflexes. A Babinski response was present on the left side. The neck was moderately stiff on passive flexion. There was tenderness on palpation of the spinous processes of the upper cervical vertebrae. Sensory examination revealed a bilateral loss of light touch, temperature, and proprioception below the level of the C-2 dermatome. There was complete loss of pain perception bilaterally from the C-2 to C-7 dermatomes. Below the level of the C-7 dermatome there was slight appreciation of painful stimuli.

Respirations were entirely abdominal in character, and the patient was moderately dyspneic. Swallowing was unimpaired, but phonation was hoarse. There was no papilledema.

A lumbar puncture performed shortly after admission showed clear and xanthochromic fluid with an opening pressure of 120 mm H2O. A Queckenstedt test showed a complete manometric block. There were 12 cells per mm3, all mononuclear, and the cerebrospinal fluid protein was 1200 mg%. Routine blood and urine studies were within normal limits.

X-rays of the upper cervical spine, both plain films and laminograms, showed erosive change in the atlanto-occipital joints on the right side; the odontoid process was tilted backward. Pantopaque cervical myelography carried out 4 days after admission showed a block at the level of C-6 (Fig. 1). The configuration of the myelographic block suggested an intradural, extramedullary tumor.

Operation. Surgery was performed 5 days after admission. Anesthesia was started under intravenously administered sodium pentothal. Immediately after induction, respiration ceased; the trachea was quickly intubated, and respiration was assisted manually by the anesthetist throughout the entire operative procedure. A midline suboccipital craniectomy and laminectomy from

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C-1 to C-7 was performed in the prone position. The dura mater was opened widely, and the tumor, red-gray in appearance and well-encapsulated, was immediately visualized. It was situated on the posterior aspect of the upper cervical cord, and extended from the level of the sixth cervical vertebra up to the obex. It covered the entire lower portion of the posterior medullary surface, displacing the cerebellar tonsils laterally. As soon as the tumor had been removed from this region, spontaneous respiration returned. It was totally removed, weighed 9.5 gm (Fig. 2), and was histologically a neurofibroma (Fig. 3). The wound was approximated in anatomic layers with interrupted silk sutures.

Upon recovery from anesthesia, the dyspnea was improved. On the third postoperative day, there was partial return of pain perception in the previously analgesic area. Thereafter, there was gradual improvement in both motor and sensory functions. Physiotherapy was instituted on March 9, at which time she was able to move both arms. Her improvement continued, and 8 months after operation the patient was able to walk unassisted. The neurological examination at this time revealed mild motor weakness with slight increase of deep tendon reflexes and hypesthesia in the right arm.

**Discussion**

Tumors in the region of the foramen magnum apparently extend upward or downward

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Fig. 1. Myelogram showing intradural type of block at C-6.

Fig. 2. Well-encapsulated extramedullary tumor, removed in toto. Total weight is 9.5 gm.

Fig. 3. Photomicrograph of the tumor (neurofibroma) (H. and E., X100).
with almost equal frequency.1 In the present case it was felt that the tumor originated at C-3 because of adhesions to the surface of the cord at this level. These tumors often produce either general or focal signs of an intracranial lesion as the result of either obstruction to the cerebrospinal fluid pathway or compression of nervous tissue by the tumor. In Love and Adson's series,2 10 of 23 cases showed papilledema and seven showed nystagmus; x-ray examination showed only one patient with enlargement of the foramen magnum on the left, the side of the suspected lesion. In four other cases there was positive evidence of increased intracranial pressure, such as erosion of the sella turcica, or increased convolutional markings on the skull. In our case there were no signs or symptoms of intracranial hypertension or other intracranial lesion. Plain x-rays of the cervical spine showed erosive changes in the atlas which simulated osteomyelitis. There were no changes in the pedicles of the spine.

An extramedullary tumor at the level of the foramen magnum produces a characteristic type of progressive motor weakness. The weakness begins in the ipsilateral arm, extends to the leg on the same side, and later involves the contralateral leg and arm. Our patient underwent such a progression of motor signs.

We considered the possibility that the preoperative dyspnea, as well as the apnea which occurred immediately after induction of anesthesia, was central in origin due to the compression of the respiratory centers in the medulla by the tumor. It was impressive that, coincident with removal of the tumor from the posterior surface of the medulla, spontaneous respiration began and continued throughout the remainder of the operative procedure.

Summary

We have reported a case of spinocranial neurofibroma in which there was recovery from flaccid quadriplegia and dyspnea after total removal of the tumor which covered the cervical cord from the level of C-6 to the obex of the medulla. Spontaneous respiration ceased abruptly as anesthesia began and returned coincident with removal of the tumor.

References