TUMORS IN THE REGION OF THE FORAMEN MAGNUM

LEON COHEN, M.D., AND DONALD MACRAE, M.D.
Department of Neurology, University of California School of Medicine, San Francisco, California

(Received for publication December 19, 1961)

Clinical neurology has established certain rules for a spinal-cord tumor to follow: (a) Long-tract signs indicate which areas of the cord are involved; focal signs, such as wasting, depressed reflexes, and segmental sensory changes, give the cross reference and pin point the anatomic site. (b) An anatomic diagnosis established, the history of the disorder should be one of steady progression. (c) There should be a partial or complete block to Queckenstedt's maneuver; and a myelogram should reveal the site.

Unfortunately, extramedullary tumors in the region of the foramen magnum are notable exceptions to the rules and thus are misdiagnosed frequently. They have been reported on by Elsberg,14 and further documented by Cadwalader,9 Frazier and Spiller,17 Stookey,27 and Globus and Strauss,29 and later summarized by Bogorodinsky,5 Symonds and Meadows,40 and Cushing and Eisenhardt.11

In the present communication 4 cases are reported and help to illustrate: (a) how the anatomic diagnosis may be erroneous, with wasting of muscles of a lower motor-neuron type occurring several segments below the level of the lesion; (b) how bizarre deficits and a relapsing, remitting course frequently lead to a diagnosis of multiple sclerosis; and (c) how myelography may be apparently normal on one or more occasions.

The appearance of atrophy in groups of muscles several segments below the caudal end of the tumor is of interest not merely because it may lead to a false anatomic diagnosis, but its causation still remains in dispute and merits review.

CASE REPORTS

Case 1. A 23-year-old male gave a 5-year history of almost constant posterior cervical pain. Fifteen months prior to admission, he had noted transient tingling along the medial aspect of his left arm when stooping or flexing his neck. One month later, a persistent numbness of the left thumb and index finger appeared, which gradually spread to involve the entire hand and all the fingers. Within 1 year he was aware of a slowness in walking, and his left knee buckled occasionally. The left hand grip became mildly weak and clumsy, and he had urinary urgency. A pneumoencephalogram was reported as normal; but it did not show air in the cervical canal. He was referred to the neurological service with a diagnosis of multiple sclerosis.

Neurological examination revealed no deficits of psychological function or of cranial nerves. There was mild but definite atrophy of all groups of muscles of the left hand, of the forearm, especially the flexor group, and of the biceps. There was mild generalized weakness of all movements of this extremity. The left calf measured 4 cm. less in girth than the right. All deep tendon reflexes were hyperactive on the left, and the left plantar response was extensor. The superficial abdominal reflexes were absent. There were dysalgiesia and hypalgiesia confined to the left C6 dermatome. Sensations of vibration and position were diminished markedly in the left upper and lower limbs, and decreased moderately in the right lower extremity. Sense of temperature was normal.

Cerebrospinal fluid manometrics were normal; the fluid contained 71 mg. per cent protein.

An electromyogram showed fibrillatory action potentials in the left 1st dorsal intersosseous muscle, confirming the clinical impression of denervation of muscles at the C8-T1 level.

A myelogram revealed the presence of an extramedullary mass at the level of the 2nd and 3rd cervical vertebrae.

Operation. A neurilemmoma was found arising from the left C3 root, which did not extend below the lower border of the C3 vertebra.

The patient made an excellent postoperative recovery.

Comment. The wasting of the left forelimb in Case 1 suggested involvement of anterior horn cells from C5 to T1, supported by an objective C6 sensory lesion, and a history
of paresthesiae of the left hand. The hyperreflexia, including the biceps reflex, indicated pyramidal involvement above C5; and this was the only correct part of the anatomic diagnosis. The location of the tumor leaves unexplained the segmental changes below C5.

Case 2. A male, aged 48 years, gave a history of numbness of the right hand of 2½ years' duration, followed within a few months by a similar numbness in the left hand. The numbness spread slowly to the shoulders. A cervical myelogram performed at the time was reported as normal. The cerebrospinal fluid contained 64 mg. per cent protein. At about this time the patient noted dyesthesiae and numbness over the inner part of the left thigh and later over the entire left lower extremity. Two months before admission, he had a similar sensory involvement of the right leg, and a second myelogram was reported as normal. A diagnosis of multiple sclerosis was entertained.

Neurological examination on admission revealed slight atrophy of the intrinsic muscles of both hands, with moderate weakness of all extremities. All deep tendinous reflexes were increased, right more than left, the superficial abdominal reflexes were absent, and both plantar responses were extensor. There were hypesthesiae over both hands, the right forearm, and the left thigh and leg, with vague borders, accompanied by dyesthesiae. There was hypesthesia over a small area of the left C4 dermatome. Sense of position was decreased in the fingers and toes, and sense of vibration was absent at the left ankle.

Roentgenograms of the cervical spine showed minimal widening of the canal at C1 to C2. Lumbar puncture demonstrated normal manometrics and the cerebrospinal fluid contained 64 mg. per cent protein. An air myelogram showed the presence of a tumor at the level of C2 without a block (Fig. 1).

Operation. An encapsulated neurofibrosarcoma, 3 \times 5 \text{ cm.} in size, was found in the interlaminar space on the right, arising from the C2 nerve root. The tumor was removed entirely.

The patient showed steady postoperative improvement in both his sensory and motor symptoms.

Comment. The sensory history and examination suggested an anatomic diagnosis at C3, or multiple lesions. The wasting of the muscles of the hand was significant and suggested a T1 lesion. The two "normal" myelograms appeared to exclude a tumor but, presumably to prevent spilling the contrast medium into the posterior fossa, were not carried higher than the C3 level. On the present occasion, air was selected for the contrast material as a tumor of the foramen magnum was suspected. The location of the tumor was consistent with the sensory and motor changes, except for the wasting of the intrinsic muscles of the hand. While the cervical roentgenogram suggested a widening of the canal, this was recognized only after air myelography had confirmed the situation of the tumor.

Case 3. A 31-year-old female began to suffer stiffness of the neck 5½ years prior to admission, followed shortly by frequent headaches in the vertex, at times lasting 7 days. Two years later she noted numbness in the ball of the left index finger and thumb, as well as paresthesiae of both feet. Some months later, she experienced ascending numbness of the left upper extremity, which spread to involve the shoulder and the left anterior area of the chest and neck to the angle of the jaw. The numbness gradually disappeared (except for brief episodes involving her shoulders) during the next 1½ years. Two years before admission, she first consulted a neurologist.

An electroencephalogram and cervical myelogram were reported as normal, but her cerebrospinal fluid contained 78 mg. per cent protein. Six months later, she first noted bilateral weakness of the leg and numbness in the right upper extremity, followed by slowly progressive weakness of both upper extremities, as well as urinary urgency and hesitancy. One year before admission she had "measles" and virtually complete quadriplegia developed rapidly, with only the left hand relatively spared. After 6 weeks a remission began and she improved to the point of being able to do all her housework, other than ironing. Two