UNUSUAL BENIGN TUMOR AT THE FORAMEN MAGNUM

REPORT OF A CASE

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It has been reported that 30 per cent of all neoplasms occurring at the foramen magnum are benign and extramedullary. In 1956, Dodge et al. analyzed 30 benign tumors situated at the foramen magnum encountered at the Mayo Clinic between 1924 and 1956, inclusive. Of these 30 tumors, 26 were meningiomas and 4 were neurofibromas, tumor types which agree in general with those reported previously in the literature. The case pertinent to this communication involves an encapsulated, extramedullary cavernous hemangioma which was extirpated completely from the posterolateral aspect of the medullospinal junction, resulting in rapid improvement of the patient's neurologic status.

The clinical history and neurologic sequelae of tumors situated at the foramen magnum have been described adequately in the literature, and it is necessary only to emphasize that this particular group of lesions, like other compressive and obstructive lesions occurring at the foramen magnum, frequently are overlooked and escape definite treatment until relatively late in the course of the disease, mainly because of the bizarre nature in which the symptoms and signs develop. The lesion in our case was no exception, for when the patient was first seen a correct diagnosis was not made, in spite of a complete neurologic investigation, including myelography.

REPORT OF A CASE

A 66-year-old man first registered at the Mayo Clinic on Aug. 11, 1956, having been referred by his home physician because of persistent paresthesias and pain affecting his left extremities. He said he had enjoyed excellent health until about 6 or 7 months before the examination in August, 1956. At the onset of this illness he first complained of a dull, aching pain in the right side of the neck, between the scapula and the occiput, extending into the supraclavicular region. The pain, which occurred daily, would begin at no particular time, but tended to be more severe in the early morning and late afternoon hours. It was aggravated by sitting, by extension of his head, as in looking up, and by exertion. The pain could be relieved by aspirin, by the application of local heat to the muscles of the neck, and by flexion of the head upon the chest. On July 11, 1956, or a month before the patient's registration, he noticed the gradual onset of a burning, tingling sensation in the left foot, which for 3 to 4 days extended up the left leg to the region of the knee and was accompanied by a sensation of coldness in the left leg to the region of the knee and in the left hand and arm. At first these sensory symptoms would come and go, but were increased by walking, and relieved by lying down. There was an impairment of appreciation and discrimination of temperature in the involved areas, described by the patient as a difficulty in distinguishing heat from cold and as a painful sensation elicited by the application of hot water to the limbs. Although

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he said he did not have loss of muscle power, he added that at times he did not seem to have full control over the movements of the left leg. As the symptoms became more persistent and constant, his home physician referred him to the Mayo Clinic for further studies.

We found the patient to have an asymptomatic abdominal aortic aneurysm for which, under other circumstances, the patient would have been advised to undergo repair. Blood pressure was 146 mm. of mercury systolic and 96 mm. diastolic. Pulse rate was 84 per minute, with regular rhythm. Auscultation of the heart revealed a grade 2 diastolic, early systolic, localized apical murmur. Results of routine laboratory studies, including urinalysis, complete blood counts and serologic tests, were normal. The erythrocyte sedimentation rate was 40 mm. in 1 hour (Westergren). Roentgenograms disclosed calcification of the thoracic aorta. Roentgenograms of the cervical part of the spinal column demonstrated narrowing of the 3rd, 5th and 6th vertebral interspaces, with hypertrophic changes that encroached upon the respective foramina. Results of examinations of the eyes, including the ocular fundi, were normal.

On Sept. 14, 1956, the patient was seen in a section of neurology. The pertinent observations made there were increased biceps and brachioradialis tendon reflex of the right arm; minimal-to-moderate loss of strength of the right serratus anterior muscle; dissociated reduction of cutaneous sensibility restricted to the left side of the body; sensation of touch appreciated normally over the entire body surface, whereas sensations of pain and temperature both were graded minus 1 to minus 2 (on a scale of 0 to +4) in the left lower extremity, and to a lesser extent over the trunk and upper extremities. In the latter area thermal sensation alone was depressed and that only minimally (graded −1).

The summary of these neurologic findings was recorded as an incomplete Brown-Séquard syndrome with an indefinite upper sensory level, suggesting, however, the presence of an intraspinal tumor at about the 5th cervical segment of the spinal cord.

As a result of these impressions cervical myelography was advised and accepted by the patient. Accordingly, on Sept. 29, 1956, lumbar puncture was done. No obstruction to the flow of cerebrospinal fluid was demonstrable by bilateral compression of the jugular veins. Cervical myelography was done with a contrast medium, iophendylate (Pantopaque). No tumor was found. Minor bony ridings at the narrowed interspaces were demonstrable, but these were regarded as insufficient to cause the patient's symptoms.

The sample of cerebrospinal fluid removed at the time of lumbar puncture was noted to be clear. It contained 40 mg. of protein per 100 cc. and 4 erythrocytes per c.mm.

The patient subsequently was dismissed to go to his home. The neurologic diagnosis was uncertain but because of the aortic aneurysm, the patient was tentatively considered to have occlusive disease of the anterior spinal artery. He was advised to return in 3 months for another examination and for consideration of the aortic aneurysm.

On Nov. 28, 1956, 2 months after he had been dismissed, the patient returned to the clinic because of progression of his symptoms. He said that from a week to 2 weeks after his dismissal he had begun to notice progressive weakness and clumsiness of his right extremities. These symptoms began in the right thumb and forefinger, causing him to drop articles. Later his gait became unsteady and unsure. Pain in the neck increased, especially in the early morning hours. A coughing and sneezing effect was marked.

During neurologic examination at the time of this second visit the Brown-Séquard syndrome was noted to be nearly complete. Loss of strength of the muscles of the right calf was graded 2 to 3; loss of strength of the muscles of the right hand and forearm, 1 to 2. Dissociated anesthesia of the left side was more pronounced; appreciation of pain and temperature below the midecervical dermatomes was now almost completely lacking. Evidence of posterior-column involvement also was present. Joint and vibratory sense was slightly impaired, especially in the left leg and right hand. Deep tendon reflex changes were similar to those noted at the previous examination; however, a marked Babinski response could be elicited in the right foot. The patient walked with a wide base and obviously was ataxic, especially on attempting to turn. Motion of the neck was limited in all directions, but was most noticeable
on flexion to the right side. Because of the marked neurologic changes, surgical exploration of the upper part of the cervical segment of the spinal cord was advised.

On Nov. 29, 1956, with the patient in the sitting position and under the influence of general anesthesia, laminectomy of the 2nd, 3rd and 4th cervical vertebrae was performed. No tumor was found. When the dura mater was opened the spinal cord appeared to be normal in all respects. However, under the 1st cervical lamina and extending into the foramen magnum was a purplish tumor (Fig. 1). Therefore, the 1st cervical lamina was removed and small suboccipital craniectomy was accomplished. The dural incision was extended up and into the posterior fossa, to a level above the tumor, which was completely intradural and unattached to the dura mater. The center of the tumor, which was round and 1.5 cm. in diameter, was at the foramen magnum. It was located on the right posterolateral aspect of the medullospinal junction and on the inferior aspect of the right cerebellar hemisphere, where it compressed the former structure to the left and the latter structure upward. It had not penetrated the pia mater; it was completely extramedullary. The tumor was lying upon the spinal accessory nerve, the spinal portion of which appeared to enter the substance of the lesion, and which subsequently was sacrificed during the extirpation. Many blood vessels entered the tumor, which obviously was very vascular; however, the bleeding was readily controlled and the tumor was delivered in its entirety without difficulty (Fig. 2). The dura mater was approximated and the wound was closed in anatomic layers.

The patient withstood the procedure well and convalescence was without incident. He was dismissed on the 10th postoperative day. At that time he was walking with minimal difficulty. Motions of the neck were normal, and the dissociated left hemianesthesia had all but abated.

By the latter part of February, 1957, the patient had recovered sufficiently to warrant consideration of surgical exploration of the abdominal aortic aneurysm; consequently, he returned to the clinic. On February 26, Dr. John W. Kirklin, of the Section of Surgery, removed a moderate-size atherosclerotic aneurysm of the lower abdominal aorta, re-establishing continuity of the vessel with a freeze-dried homologous aortic graft. Again the patient made an uneventful recovery.
Neurologic examination at the time of this visit demonstrated further improvement. The patient had no complaints of neurologic origin, although upon careful testing it was evident that he was still somewhat ataxic when he attempted to walk in tandem. However, in other respects the patient was normal.

COMMENT

In 1945 Noran published detailed descriptions of all intracranial vascular tumors and malformations. He aptly emphasized at that time the confusion that existed in the literature in regard to these lesions. He summarized eight of the classifications commonly used in the world literature and added one of his own. It can be seen from his paper, as well as from a review of the original contributions, that the lesion generally referred to as a "cavernoma" or "cavernous hemangioma" is variously grouped with the neoplasms or the malformations or by itself.

Fig. 3. Section of the benign cavernoma (hematoxylin and eosin; ×50).

It is now generally conceded that cavernomas are produced in the embryonic stage of development and are formed from vessels that have reached maturity. Grossly, these lesions are most often found within the substance of the brain, and are described as consisting of closely packed, thin-walled vessels of varying sizes supported on a framework of a reticular connective tissue. The lesions are soft and compressible, like a sponge. Within the substance of the brain they are usually sharply demarcated, although at times some vessels may extend out into the nervous parenchyma. Since these lesions rarely are the sources of intracranial bleeding, they are found infrequently at operation. Indeed, Cushing reported only one instance of a cavernoma in a series of 1,522 histologically verified intracranial tumors.

The tumor in our case consisted of many thin-walled vascular spaces which were lined by simple endothelium overlying a layer of hyalinized connective tissue (Fig. 3). Considerable collagenous connective tissue was found throughout the stroma of
the lesion, which was relatively acellular and in places showed evidence of degeneration. Condensation of the stroma at the surface constituted the "capsule" of the malformation. The vessels were found in small groups and clusters with leukocytic infiltration about these zones. van Gieson's stain for elastic tissue and Mallory's phosphotungstic acid-hematoxylin stain were used, and hematoxylin and eosin stain was employed routinely. No elastic fibers were found within the walls of the blood vessels, and there was no evidence that any glial or meningeal elements were present. The lesion was not an angioblastic meningioma. Indeed, there were no areas in which active cellular proliferation could be seen. The lesion unquestionably was a cavernous hemangioma with somewhat more connective-tissue stroma than is usually encountered in these lesions when they are found within the substance of the brain.

SUMMARY

A case in which an unusual benign tumor developed at the foramen magnum in a 66-year-old man is presented. The lesion, which was completely extirpated, proved microscopically to be a cavernous hemangioma. It was extramedullary and intradural in position. Clinically, the tumor was indistinguishable from the commoner true neoplasms found at this site. Brief reference is made to the perplexing literature on intracranial vascular tumors and malformations.

REFERENCES