Extradural hemangioblastoma of the spinal cord

Case report

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A case of hemangioblastoma of the spinal cord is described in which the tumor presented in an intradural extradural location. The patient did not have von Hippel-Lindau's disease or metastatic seeding. It is assumed that the tumor arose in the pia, and its direction of growth was purely extradural.

KEY WORDS
hemangioblastoma • spinal cord neoplasm • von Hippel-Lindau's disease

HEMANGIOBLASTOMA of the spinal cord is a rare tumor; its incidence varies from 1.6% to 3% of primary spinal cord tumors. The tumor presents most frequently as a single intramedullary neoplasm. It is more common in men. The association with von Hippel-Lindau's disease is well known and occurs in 30% of cases. Polycythemia has never been reported with a purely spinal hemangioblastoma, although Yaşargil, et al., recently stated that it "... is present in 9% of cerebellar hemangioblastomas and a similar figure with spinal lesions is probable." Clinical significant intradural extramedullary hemangioblastomas are extremely rare. Wyburn-Mason reported five surgically verified hemangioblastomas arising from spinal roots (his Cases 49, 50, 51, 58, and 60), although his Case 58 "... secondarily invaded the cord." It is not unusual, as judged from published reports of careful autopsies done on patients with von Hippel-Lindau's disease, that clinically insignificant and minuscule hemangioblastomas are found on spinal roots or attached to the pia mater.

The present report is prompted by the rarity of a hemangioblastoma presenting as an intradural extradural tumor.

Case Report

This 59-year-old man was admitted after 2 years of low-back pain and 1 year of slowly progressive gait disturbance. For 6 months he had noticed a sensation of numbness over the right buttock and posterior thigh.

There was no familial history of von Hippel-Lindau's disease. A twin sister was normal. An older sister had died at the age of 55 years of a ruptured intracranial aneurysm.

Examination. The patient had a limp on the left side with superimposed mildly spastic gait, and weakness of dorsiflexion of the left
foot. There was atrophy of the left gluteal and quadriceps muscles. The left knee jerk was absent, and the right depressed; the left ankle jerk was hyperactive, and the right depressed. There were no Babinski signs. Sensory testing disclosed hypalgesia to the T-12 level on the left. There was a single 2 × 3 cm café-au-lait spot over the left side of the neck.

Routine laboratory studies were normal; there was no polycythemia. An intravenous pyelogram performed at the referring hospital was normal. Plain spine x-ray films showed slight thinning of the right T-12 pedicle. The cerebrospinal fluid was deeply xanthochromic, clotted spontaneously, and contained more than 2000 mg/dl protein. Lumbar myelography showed a complete block opposite the T-12 level, and the appearance was that of an intradural extramedullary tumor on the left side displacing the cord.

Operation. The tumor was exposed by a laminectomy covering T-11 through L-1. The bone and extradural tissues were normal. The dura was opened in the midline and a spherical purplish tumor was found attached to the left posterior aspect of the spinal cord, covered by arachnoid (Fig. 1 left). A single large tortuous vein was present superiorly, and several tortuous smaller vessels were intermingled with the roots of the cauda equina inferiorly. There was no dural attachment. The tumor was cleanly separated from the surface of the spinal cord by microdissection, and fine bridging vessels between the cord and the tumor were divided. The posterior root of T-12 was intimately adherent to the anterior surface of the tumor, and was sacrificed during removal (Fig. 1 right). A computerized tomogram of the head performed postoperatively was normal.

The patient's recovery was uncomplicated. He returned to work 6 weeks postoperatively, and has remained asymptomatic during a 10-month follow-up period.

Pathologicai Examination. The specimen was a 3 × 2 × 1 cm dark red solid mass partially covered by a thin fibrous membrane.
Extramedullary hemangioblastoma of spinal cord

The cut surface was spongy with small blood-filled cysts and yellow areas.

Microscopically, the tumor consisted of vascular channels, separated by an intervascular stroma with a variable cell density and collagenous tissue (Fig. 2). The vascular spaces ranged in size from capillaries to veins and were lined by flattened endothelial cells; the walls varied in thickness. The stromal cell nuclei appeared uniformly benign. The amount and appearance of the eosinophilic cytoplasm of the stromal cells ranged from scanty to moderate. Some cells had numerous long cytoplasmic processes that connected with those of other cells, and others were vacuolated and contained sudanophilic material. The capsule consisted of collagenous tissue, which in one area had peripheral nerve fibers.

Discussion

Hurth, 3 in an extensive review of the literature (138 cases), found 38 hemangioblastomas that were intradural extramedullary in location; 28 were located on nerve roots (16 in the cauda equina, 10 on posterior roots and two on anterior roots). Ten were extramedullary in location, as in the current report. Over 50% of the tumors were in patients with the classical stigmata of von Hippel-Lindau's disease; approximately 50% were asymptomatic and incidental findings at autopsy. Simultaneous separate intradural spinal tumors were found in multiple combinations (such as intramedullary plus extramedullary/root, and extramedullary plus root) in several patients.

Hemangioblastomas arise most commonly in the posterior columns of the spinal cord, or extend from the pia on the posterior surface of the cord into the posterior columns. 6,7,9,10 Rubinstein 6 has observed that this tumor always reaches the pia in the cerebellum and spinal cord.

It appears that the direction of growth of a hemangioblastoma, assuming its origin in the pia, determines its ultimate relationship to the spinal medulla. Intramedullary tumors with an extramedullary excrecence have been described by Wyburn-Mason 11 and Hurth. 3 In our patient, who had no evidence of von Hippel-Lindau's disease, a hemangioblastoma arose in the pia mater of the spinal cord. Its direction of growth was purely extramedullary. There was no clinical or labora-

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References


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