Intramedullary hemorrhage in spinal cord hemangioblastoma

Report of two cases

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The authors describe two cases of intramedullary hemorrhage caused by thoracic hemangioblastoma. Both patients presented with acute paraplegia. The lesion in the first case was diagnosed by myelography and in the second by magnetic resonance imaging. Emergency surgical evacuation of the intramedullary hematoma and tumor was performed in these patients. Hemangioblastoma was confirmed by histopathological examination in both cases. Both patients remain paraplegic after 7 and 1 years, respectively. Intramedullary hemorrhage is a rare and devastating effect of spinal hemangioblastoma.

KEY WORDS • hemangioblastoma • intramedullary hemorrhage • subarachnoid hemorrhage • spinal cord tumor • von Hippel-Lindau disease

H EMANGIOBLASTOMA of the spinal cord occurs either sporadically or in association with von Hippel-Lindau disease, an autosomal-dominant inherited disorder manifested by tumors in several organs. Patients usually present with slowly progressive paraparesis.11 Hemorrhage secondary to spinal hemangioblastoma is quite rare. Only seven cases have been reported and all involved subarachnoid hemorrhage (SAH), not intramedullary hemorrhage.2–4,10 Five hemorrhages were in the cervical spinal cord, one was in the thoracic area, and one was associated with a lumbar nerve root. None of these cases presented with myelopathy; however, the cervical cases displayed symptoms similar to those of SAH from an intracranial lesion.2,4,10 Surgical removal of the tumor left patients neurologically normal,2,4 or with some upper-extremity weakness.10

We present two cases of intramedullary hemorrhage from thoracic spinal hemangioblastomas, causing acute paraplegia. Both patients were operated on immediately and, at follow-up examination, remain paraplegic. These cases illustrate one aspect of the natural history of spinal cord hemangioblastomas and argue for more extensive diagnostic screening of patients with von Hippel-Lindau disease. Although rare, intramedullary hemorrhage must be considered a possible complication of spinal hemangioblastoma.

Asymptomatic spinal hemangioblastoma in patients with von Hippel-Lindau disease syndrome should be followed with this potential complication in mind. Conversely, hemangioblastoma must be considered among the differential diagnoses for a patient presenting with an intramedullary hemorrhage.

Case Reports

Case 1

This 31-year-old previously healthy woman noted abdominal cramping that radiated posteriorly 2 days prior to admission. The next day, she reported burning dysesthesias over the upper back and shoulders. On the day of admission, the patient developed neck pain and, within 1 hour, developed numbness below the upper chest and inability to move her legs.

Examination. On physical examination, there were no stigmata of von Hippel-Lindau disease. The patient was awake and alert but had complete sensory loss below C-8 on the left side and C-7 on the right. There was decreased sensation to pinprick over the medial left hand, forearm, and axilla. There was weakness of the intrinsic hand muscles of the left hand. The lower-extremity deep tendon reflexes were absent and Babinski signs were present bilaterally. A lumbar
myelogram revealed a complete block at the T6–7 level secondary to an intramedullary mass. There were 3060 red blood cells per cubic millimeter and one white blood cell per cubic millimeter in the pink cerebrospinal fluid.

**Operations.** The patient was immediately taken to the operating room. A laminectomy of the T3–6 level was performed. The dura was tense and nonpulsatile. It was opened longitudinally in the midline for the full extent of the exposure. There were several prominent serpiginous veins on the posterior aspect of the pale distended spinal cord. A midline myelotomy was made over the midportion of the exposed cord. At a depth of 2 to 3 mm a hematoma associated with a vascular mass was encountered; both were microsurgically excised. The tumor was 5 cm in length; pathological examination confirmed the diagnosis of hemangioblastoma. Postoperatively, the patient’s thoracic myelopathy was unchanged. Seven years later, she has a complete thoracic myelopathy at T-2.

**Postoperative Course.** She is now spastic in the lower extremities, but has normal strength and sensation in the upper extremities. A follow-up magnetic resonance (MR) image revealed no intracranial lesions, myelomalacia in the upper thoracic cord, and a small syrinx in the upper cervical cord.

**Case 2**

This 28-year-old man had been diagnosed with von Hippel-Lindau disease 11 years earlier. During sexual intercourse, he experienced an electric shocklike sensation in his lower back, which radiated to his groin, his buttocks, and down his legs to his feet. Paresthesias progressing to numbness began on the soles of his feet and gradually ascended to his thorax. Upon presentation at a local hospital, he was paraparetic but still able to lift his legs off the bed. Sensation below T-8, including the perineal region, was absent.

**Examination.** After transfer to Massachusetts General Hospital 2 hours later, his mental status, cranial nerve function, and arm strength were normal; the lower extremities were hypotonic but not flaccid and immobile. There was complete loss of sensation to pinprick, light touch, temperature, vibration, and proprioception below T-7 on the right side and T-6 on the left. Only the upper-left abdominal reflex was present, patellar reflexes were absent, ankle reflexes were 2+ bilaterally, and the toes were downgoing. The bulbocavernous reflex was absent, and anal tone was decreased. A gadolinium-enhanced MR image of the spine revealed a mass at T-7 level (Fig. 1).

**Operation.** The patient was immediately taken to the operating room where a T7–8 thoracic laminectomy was performed. The dura was taut and as it was opened, a large, red, fungating intramedullary mass with serpiginous feeding artery and draining vein was seen (Fig. 2). The mass contained an intramedullary hematoma at the inferior pole of a hemorrhagic tumor. Microsurgical evacuation of the hematoma and resection of the tumor were accomplished. Pathological examination confirmed the diagnosis of hemangioblastoma (Fig. 3).

**Postoperative Course.** Follow-up examination at 6 months revealed continued spastic paraplegia and loss of sensation below the umbilicus. Urodynamic studies documented a spastic bladder.

**History.** The diagnosis of von Hippel-Lindau disease had been made when the patient was 17 years of age and presented with retinal hemangiomas. Renal cell carcinoma status was confirmed after a left nephrectomy in 1990, and a computerized tomographic scan of the head was negative. There is an extensive family history of von Hippel-Lindau disease including his mother, sister, brother, two uncles, several cousins, and one nephew.

**Discussion**

**Clinical Presentation**

Thirteen percent of all hemangioblastomas occur in the spinal canal. Most cases present with spinal cord compression that worsens over several years. When
SAH occurs due to hemangioblastoma, symptoms are similar to those of SAH secondary to an intracranial lesion, except that spinal cord or root deficits may be detected. The two cases we present differ from the previously reported cases of hemorrhagic spinal hemangioblastoma in that a rapidly progressive paraplegia developed secondary to a catastrophic intramedullary hemorrhage.

**Diagnosis**

The MR images in the second case allowed definitive diagnosis prior to surgical treatment and showed the precise relationships among tumor, hematoma, spinal cord, and dura. In the first case, preoperative diagnosis was provided by angiography and myelography, which are less informative and more invasive than MR imaging.

**Surgical Management**

Using microsurgical operative techniques, complete removal of spinal hemangioblastoma with low morbidity is possible. The two patients presented here underwent emergency surgery because of progressive paraplegia. Both patients were paraplegic by the time of surgery, and there has been no recovery of lower-extremity function, although upper-extremity paresis improved in the first case. The rapid progression of their paraplegia prevented intervention prior to the development of a complete deficit.

The role of early operation of intramedullary hematoma secondary to tumor is undetermined because of the paucity of cases reported and limited follow-up review. There are two reports of deaths within 2 weeks of presentation of a large spinal SAH secondary to tumor. There are also reports of improvement of acute cauda equina syndrome and incomplete myelopathy after early surgery for SAH secondary to spinal tumors. More cases must be evaluated to determine the role of surgical therapy for patients with complete myelopathy. The two cases presented suggest a dismal prognosis for motor improvement from paraplegia.

Subarachnoid hemorrhage of the spinal cord is usually secondary to an arteriovenous malformation. Spinal cord tumors are the second most likely etiology, ependymomas accounting for over 80% of these. Spinal hemangioblastoma presents less frequently with SAH although half the cases of spinal hemangioblastoma in one review presented with SAH. Most of these patients had minor neurological deficits; however, as evident in these two cases, neurological catastrophe can occur. Early operation at the first presentation of neurological deficit may avoid the rapid deterioration associated with intramedullary hemorrhage. Given the low morbidity of microsurgical resection and the high incidence of SAH, we recommend early surgical removal of spinal hemangioblastomas that have bled.

As outlined by Neumann, et al., discovery of a hemangioblastoma warrants evaluation for von Hippel-Lindau disease, because 23% of patients with hemangioblastoma have this disease. The recent iden-
Identification of the von Hippel-Lindau disease tumor suppressor gene will allow screening of family members to facilitate accurate diagnosis before presentation of the disease. Because intramedullary hemorrhage is a potential, if rare, presentation of spinal hemangioblastoma in a patient with or without von Hippel-Lindau disease, we would continue to follow these cases.

References

Manuscript received January 4, 1994. Accepted in final form February 9, 1994.
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