Spinal intramedullary metastatic medulloblastoma

Case report

B. JOSEPH ZUMPANO, M.D.
Department of Neurosurgery, Dartmouth Medical School, Dartmouth-Hitchcock Medical Center, Hanover, New Hampshire

Metastatic spread of medulloblastoma along the neuraxis by leptomeningeal seeding through the cerebrospinal fluid pathways is well known. The occurrence of extracranial metastases outside the neuraxis has been well established, but the occurrence of intramedullary spinal cord metastases not related to surface seeding is rare. A histologically documented case of the latter type is described.

KEY WORDS • metastatic medulloblastoma • spinal intramedullary metastasis

The occurrence of spinal intramedullary metastases in the absence of surface leptomeningeal seeding is a most unusual metastatic presentation for a cerebellar medulloblastoma. Such a case is reported, in which the patient presented with the myelographic appearance of an intramedullary spinal cord tumor about 13½ months after the removal of his primary cerebellar tumor.

Case Report

This 8-year-old boy was admitted to the Dartmouth-Hitchcock Medical Center on October 1, 1975, with a 3-week history of headache and ataxia. His schoolteacher reported a progressively poorer performance in his schoolwork for about 1 year. An ophthalmologist found florid papilledema and referred the child for neurosurgical evaluation.

First Admission. Physical findings consisted of a left sixth cranial nerve palsy, severe bilateral papilledema, and truncal ataxia. A posterior fossa arteriogram and ventriculogram confirmed the presence of a large left cerebellar mass lesion and hydrocephalus. Cerebrospinal fluid protein was 96 mg%. A posterior fossa craniotomy was performed on October 7, 1975, at which time a left cerebellar tumor was excised. Histology verified the presence of a medulloblastoma.

Over the following 2 months, the patient was given radiation treatment totaling 3500 rads to the brain and 3420 rads to the spinal cord. His hydrocephalus remained unchecked and required a ventriculoperitoneal shunt with two subsequent shunt revisions. He was seen in follow-up on February 18, 1976, at which time he was without ataxia, had normal fundi, and had returned to school. The remainder of his neurological examination was normal. He was seen again on October...
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13, 1976, at which time the fundi were still normal; he appeared to have a very mild generalized ataxia, slightly more pronounced on the left side.

Second Admission. The patient was readmitted on November 26, 1976, with a 2-week history of progressive weakness of his left arm with inability to abduct it. There was no headache, diplopia, or ataxia; but he did complain of posterior neck pain with hyperextension.

On physical examination, the cranial nerves were intact. Fundoscopic and extraocular muscle examinations were normal. There was no hypotonia, nystagmus, or dysmetria, but a mild persisting truncal ataxia was noted. Hyperextension of the neck produced posterior neck pain without a Lhermitte's phenomenon. There were hyperactive deep tendon reflexes in the lower extremities with bilateral Babinski responses. Biceps jerks were absent. There was weakness of the biceps, deltoid, supraspinatus, infraspinatus, and triceps, more pronounced on the left than on the right side. A sensory level could not be demonstrated, and proprioceptive testing was considered unreliable due to the child's poor cooperation.

A cervical myelogram showed an almost complete block to flow of the contrast material at the C-4 and C-5 levels, although the spinal cord appeared widened from C-2 to C-7. Cerebrospinal fluid protein was 416 mg%. A computerized tomographic (CT) scan of the brain failed to reveal the presence of intracranial recurrence or spread of tumor. A CT scan of the high and mid-cervical regions revealed the presence of an eccentrically-placed, low-density cavitary lesion to the left of center within the white matter of the spinal cord (Fig. 1). Two nodular areas of increased density could be seen ventral to the cavitary lesion.

Operation. Exploration of the cervical spinal cord was performed to determine whether the lesion was a cyst, syrinx, or a new primary spinal cord tumor. At surgery, the spinal cord was needled at three different levels, including the widest areas at about C-5. No fluid was obtained. We did not see the typical opaque leptomeningeal seedings that produce the “candle dripping” myelographic appearance of surface metastases. The spinal cord was incised along the dorsal root entry zone from the C-2 through C-6 levels. This area of the spinal cord appeared to be the most fluctuant. A cystic cavity containing a blood clot was entered at the lower one-half of our incision and was decompressed. Upon extending the incision upward above the C-5 laminar level, solid reddish-purple and gray tissue was encountered within the deep spinal cord substance. A distinct plane of dissection was not found so that multiple biopsies were taken and the procedure was then terminated. Histological examination of all biopsy specimens from the deep cord substance verified the presence of medulloblastoma.

Postoperative Course. Postoperatively, the child’s left upper extremity monoparesis failed to improve and he could walk only with assistance.

Discussion

Distant metastases from primary central nervous system tumors in children most often arise from medulloblastomas, neuroblastomas, meningeal sarcomas, and less often, ependymomas. Smith, et al., in reviewing 8000 primary central nervous system tumors of the Armed Forces Institute of Pathology, found 35 well documented cases of distant metastatic spread. He noted that medulloblastoma was second only to glioblastoma multiforme in incidence of metastases. There
were eight cases of metastatic medulloblastoma involving either bone, lymph nodes, lung, or liver. Seven of the eight cases had diffuse leptomeningeal surface seeding of the spinal cord. Others have documented well the extra-axial spread of this tumor to the lymphatics and cervical lymph nodes in particular, as well as to lung, liver, kidney, spleen and pancreas, pelvis and long bones, spine, flat bones, skull, vertebrae, ovary, parotid, bone marrow, neck muscles, and mediastinum.1,2,6,7,9

Both surgery and radiation therapy have been implicated in encouraging metastases.8 Brutschin and Culver,2 in reviewing 60 cases exhibiting extracranial metastases, noted a male-to-female ratio of 3:1, and an average survival time of 2.5 years after exploration. The mode of metastatic spread appears to be by local invasion, hematogenous dissemination, seeding of cerebrospinal fluid pathways, and less likely by lymphatic dissemination.4,6,8

Metastatic medulloblastoma appearing as an intramedullary spinal cord tumor is a rare and unusual presentation. Edelson, et al.,4 in reviewing the literature up to 1972 on intramedullary spinal cord metastases, found only 70 reported cases. Intramedullary spinal metastases represented 3.4% of 175 metastatic spinal cord lesions, 50% of which were from carcinoma of the lung; malignant melanoma and lymphoma appeared to be second and third in frequency of intramedullary metastatic spread.

Deutsch, et al.,9 recently reported three cases of metastatic medulloblastoma to the spinal cord in which two of the patients were clearly found to have intramedullary metastatic lesions by myelography. No mention was made of histological verification of the tumor other than for the presence of positive spinal fluid cytology for malignant cells in all three cases.

Guyer, et al.,9 did not note the myelographic appearance of a widened spinal cord as a metastatic presentation. They found positive spinal fluid cytology for malignant cells in 30% to 50% of malignant brain tumors, even without seeding of the leptomeninges.

The striking absence of surface seeding of the dorsal spinal cord in the presence of deep intraparenchymal metastases is an unusual finding in our patient. Histological verification of the invasive medulloblastoma diffusely spread throughout the deep white matter of the spinal cord is seen in Fig. 2. The
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typical tumor cells, consisting of dark-staining nuclei of slightly irregular shape surrounded by scant cytoplasm appeared to be histologically identical with the primary tumor.

The mechanism by which the intramedullary spread of this tumor occurred remains a matter of speculation. Whereas it is inviting to attribute its origin to spread from the cerebellum downward through the central canal, this judgment must be tempered by the observations that 1) the cystic component of the metastasis, although containing tumor in its wall, was mainly a hematomyelic cavity situated laterally and well away from the central canal, and 2) multiple biopsy specimens taken well away from the central canal attested to the invasiveness and diffuse nature of the lesion. It does, however, remain a prime consideration.

It is possible that some element of extension or invasion occurred along vascular channels, as evidenced by the frequent and extensive perivascular cuffing by tumor cells seen along the deep intraparenchymal blood vessels of the spinal cord (Fig. 2). One could postulate that blood vessel destruction and hemorrhage into the metastasis may have accounted for the hematomyelia seen in this patient, but the presence of perivascular cuffing by tumor cells does not necessarily imply the presence of a destructive process to the vascular wall. Direct tissue invasion and extension from the cerebellum probably played a major role.

Summary

A case of intramedullary spinal metastasis of a cerebellar medulloblastoma is presented. This little-recognized mode of presentation is established by histological documentation and myelography, and supported by the experience of others.a

References


Address reprint requests to: B. Joseph Zumpano, M.D., 8740 North Kendall Drive, Suite 110, Miami, Florida 33176.