Spinal intramedullary spread of medulloblastoma

Case report

STANLEY L. BARNWELL, M.D., PH.D., AND MICHAEL S. B. EDWARDS, M.D.

Departments of Neurological Surgery and Pediatrics, School of Medicine, University of California, San Francisco, California

The case of an 11-year-old boy is reported in whom two intramedullary lesions developed at the thoracic-cervical and thoracic-lumbar junctions 2-3 years after resection and irradiation of a medulloblastoma in the posterior fossa. There was no evidence of subarachnoid spread of the tumor. Magnetic resonance imaging was used to localize these lesions, and provided much better diagnostic information than either computerized tomography scans or myelograms.

KEY WORDS • magnetic resonance imaging • medulloblastoma • spinal intramedullary metastasis

While metastasis of medulloblastoma throughout the spinal subarachnoid space is a well-known phenomenon, dissemination of primary cerebral lesions down the central canal of the spinal cord to give a purely intramedullary lesion is a very unusual pathological finding. To our knowledge, Zumpano9 has reported the only other instance of intramedullary spread of medulloblastoma. Several features of Zumpano's case and our case suggest a putative mechanism for this occurrence.

Case Report

This 11-year-old boy developed headaches, nausea, and vomiting without focal neurological deficits in August, 1980. A computerized tomography (CT) scan revealed the presence of a midline posterior fossa mass. At craniotomy in September, 1980, a cerebellar vermis tumor that was shown at histopathological examination to be a medulloblastoma was resected from the posterior fossa. Postoperatively, the patient was irradiated with 5500 rads to the posterior fossa and 4500 rads to the whole brain and spinal axis. Two months after surgery, he developed a cerebrospinal fluid (CSF) leak from the surgical wound and Streptococcus salivarius meningitis, which resolved after antibiotic therapy. The fistula healed spontaneously.

The patient did well until the summer of 1983, when he developed a slowly progressive gait abnormality and occasional back pain. There was no bowel or bladder dysfunction. He was admitted to the Pediatric Neurosurgery Service for evaluation of these symptoms on March 12, 1984. He had been taking no medications.

Examination. The second through 12th cranial nerves were normal. Motor tone, bulk, and strength were normal in both arms. Sensory testing to pinprick, light touch, temperature, vibration, and proprioception was also normal in both arms. In particular, there was no dissociated sensory loss in either arms or legs. The strength in both legs was minimally decreased with an associated loss of tone. Response to pinprick, temperature, vibration, and proprioception were markedly decreased in both legs. Tests of coordination showed that the patient had normal finger-to-nose response but a poor heel-to-shin response. Deep-tendon reflexes were absent in both legs. The Romberg test was positive. His gait was wide-based and unsteady, and he could walk only with the aid of Canadian crutches.

A CT brain scan with and without a contrast agent showed the site of craniotomy and surgical resection but no signs of recurrent tumor or hydrocephalus. Electromyographic and nerve conduction velocity studies provided no evidence of a neuropathy. At an attempted pan-myelography, during which metrizamide was instilled by lumbar puncture, a complete block was found at the L-1 vertebral level. A subsequent CT scan of the cervical and thoracic spine showed the presence
Fig. 1. Sagittal magnetic resonance image of the cervical, thoracic, and lumbar spine using spin-echo and inversion recovery techniques. Two intraspinal masses can be seen, one at the cervicothoracic junction extending from approximately C-6 through T-2 and the other at the thoracolumbar junction extending from approximately T-8 through L-1. The more inferior mass completely fills the spinal canal in a cylindrical fashion (curved arrow). The upper lesion has the appearance of a spindle-shaped enlargement of the cord (straight arrow). Both lesions were compatible with a diagnosis of an intramedullary mass.

Operation. A total laminectomy of T-10 and T-11 was performed on March 16, 1984. After the dura was opened, the arachnoid was observed to be markedly thickened. Biopsy specimens taken from the arachnoid showed no evidence of tumor. A midline dorsal myelotomy was made, and necrotic tissue herniated from the central portion of the cord. No attempt was made to resect the tumor; several biopsy specimens were obtained, however, and histopathological examination of frozen sections showed the tissue to be medulloblastoma.

Postoperative Course. With the exception of transient urinary retention, there were no postoperative complications. After one course of chemotherapy with 6-thioguanine, procarbazine, dibromodulcitol, 1-(2-chloroethyl)-3-cyclohexyl-1-nitrosourea (CCNU), and vincristine, the patient was able to walk without the use of Canadian crutches. His ankle jerks returned and proprioception in both legs was improved. After three courses of chemotherapy, his leg strength improved so dramatically that he was able to play baseball. The patient has received a total of six courses of the same chemotherapy and remains neurologically stable.

Discussion

Medulloblastoma, malignant ependymal tumors, and malignant tumors of the pineal region have the greatest potential for dissemination to the neuraxis and for extraneural systemic metastases. Metastases occur either spontaneously or after shunting procedures, surgery, or radiation therapy have been performed. The malignant potential of the tumor and its proximity to the CSF circulation are necessary conditions for the metastatic spread of intracranial tumors to the spinal cord. Intracranial tumors usually spread throughout the subarachnoid space, presumably carried by the CSF circulation, with no predilection to a particular area.

Therefore, the occurrence of intramedullary metastases with no surface leptomeningeal seeding is a very unusual clinical finding. To our knowledge, there is only one other reported case. In 1978, Zumpano reported the case of a patient who developed a lesion in the spinal cord 13 months after resection of a cerebellar medulloblastoma. At laminectomy, tumor was found solely within the spinal cord parenchyma; no subarachnoid seeding was noted at the site of exploration. Zumpano suggested that the tumor had spread from the cerebellum through the central canal of the spinal cord. He noted, however, that the cystic component of the metastasis was situated far lateral to the central canal and that the tumor had spread throughout the spinal cord. Therefore, the primary site of metastasis could have been located away from the central canal of the spinal cord.

These two cases have several features in common. Both patients had hydrocephalus during the course of their illness, both had undergone resection of a primary cerebral lesion before the spinal metastatic lesion developed, and both were treated with radiation therapy after the initial resection of the posterior fossa mass. The occurrence of hydrocephalus in both patients, which could dilate the opening of the central canal to allow seeding into the cord, and the location of the primary lesion in the posterior fossa suggest that the central canal may be the pathway for spread of malignant tumors. Our case was unusual because two lesions were found. On the basis of MRI, we could not determine if there was contiguous spread or two distinct lesions.

The rarity of these lesions may in part reflect the sensitivity of radiological diagnostic techniques. Spinal
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CT scans and myelograms cannot always be used to distinguish intramedullary from extramedullary spinal cord masses, especially when an extensive subarachnoid block is present. In our patient, MRI clearly showed the central location of the two metastatic lesions, and the surgical approach was planned to allow a midline dorsal myelotomy to be performed if the tumor was not found when the dura was opened. The operative finding that the mass of necrotic tissue and tumor was centrally located and the fact that multiple biopsy specimens obtained from the surface of the spinal cord and meninges showed no evidence of tumor confirm the diagnostic value of MRI for these lesions. The availability of MRI may lead to the identification of more instances of this type of lesion.

Acknowledgments

We thank J. Fernando Elias, M.D., Gould Medical Group, Modesto, California, for referring this patient; Cindy Huff for typing the manuscript in draft; and Neil Buckley for editing the paper.

References


