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Childhood Onset of Myxopapillary Ependymomatosis: MR Features

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Summary: We report a 12-year-old girl who at presentation had diffuse myxopapillary ependymomatosis of uncertain primary site. The MR appearance and mechanism of metastases of the tumor are discussed.

Index terms: Spinal cord, neoplasms; Brain, neoplasms; Ependymoma; Children, neoplasms

Myxopapillary ependymoma is a distinct subtype of ependymoma that was defined by Kernohan (1). This tumor is essentially restricted to the filum terminale or conus medullaris (2–6). The occurrence of myxopapillary ependymoma of the spinal cord in children is relatively rare (6). We report a case presenting with diffuse subarachnoid spread of myxopapillary ependymoma of the spinal cord of uncertain primary site. The magnetic resonance (MR) appearance and histologic features of the myxopapillary ependymoma are described along with a discussion of the mechanism of metastases of the tumor.

Case Report

A 12-year-old girl presented after 2 months of progressive pain and stiffness in the neck and lower back associated with a tense and stooped-over gait. About 5 days before admission, she had noted intermittent visual blurring, headache, nausea, and vomiting. She had no paresthesia or muscle weakness. Bowel and bladder functions were normal. Physical examination was significant for tenderness to palpation over the lumbar and sacral spine. There were no meningeal signs nor any focal neurologic abnormality. The remainder of the physical examination was unremarkable. Routine laboratory studies including cerebrospinal fluid (CSF) analysis were normal.

Findings on skull and spinal radiographs were unremarkable. MR imaging was performed with a 1.5-T system. T1-weighted images of the spine obtained before and after contrast injection showed diffuse enhancement of pial membranes throughout the cervicothoracic spinal cord and in the fourth ventricle (Fig 1). Heterogeneously enhancing nodules were noted in the conus medullaris, cauda equina, and terminal thecal sac (Fig 2). MR imaging of the head showed diffuse enlargement of all ventricles consistent with a communicating hydrocephalus. Axial MR images at the level of fourth ventricle showed subarachnoid enhancement without a focal enhanced mass. The pituitary infundibulum was thickened and isointense relative to normal white matter on T1-weighted images and homogeneously hyperintense on proton density– and T2-weighted images. Postcontrast T1-weighted images showed marked enhancement of the thickened infundibulum with extension into the hypothalamus (Fig 3). Findings of serial CSF cytology, chemistry, and microbiology were normal. Further evaluations including bone marrow biopsy; whole-body technetium-99m–methylene diphosphonate bone scan; and CT scans of the head, chest, abdomen, and pelvis yielded no additional information.

After lumbar laminectomy and opening of the dura, the cauda equina was noted to be grossly abnormal with a sugarlike coating of tumor tissue on the nerve roots causing their adhesion; no solitary or encapsulated mass was identified. Excisional biopsies of the tumor tissue coating the cauda equina and the base of the thecal sac were obtained. Because of the adherent nature of the tumor and its diffuse distribution, surgical resection was not considered feasible. Resection of the infundibular mass was not attempted, however, a ventriculoperitoneal shunt was placed for relief of hydrocephalus.

Microscopically, the tumor tissue consisted of fibrillated cells with both glial and epithelial features. The cells formed glandlike spaces with numerous adjacent blood vessels exhibiting perivascular anuclear zones or "pseudorosettes." There was mild to moderate mucin accumulation between islands of tumor cells (Fig 4). Flow cytometry revealed a diploid deoxyribonucleic acid index with low S phase of less than 5%. The pathologic findings were diagnostic of myxopapillary ependymoma.

The patient underwent two courses of chemotherapy. Four months after treatment, MR examination of the brain and spine showed no interval change in the appearance of the tumor. The patient's condition continued to deteriorate, and the infundibular lesion was irradiated. In a fol-

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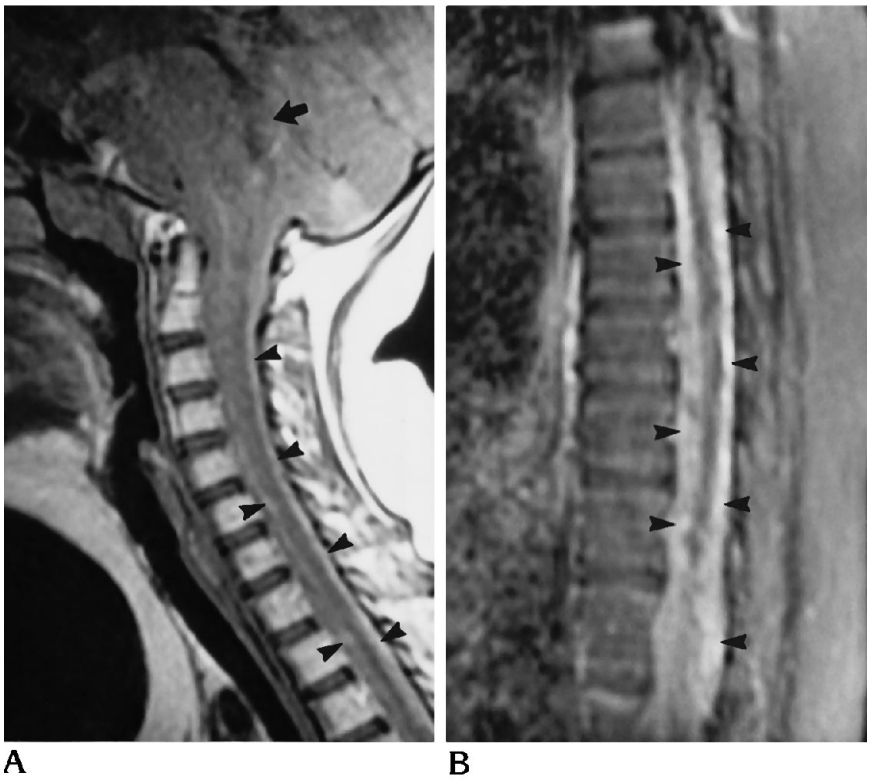

Fig 1. Contrast-enhanced sagittal T1-weighted spin-echo MR images of cervical (350/19/2 [repetition time/echo time/excitations]) (*A*) and thoracic (433/19) (*B*) spine show levels of the diffuse leptomeningeal enhancement throughout the cervicothoracic spinal cord (*arrowheads*) and CSF enhancement in the floor of the fourth ventricle (*arrow*).

low-up MR examination 6 months after diagnosis, there was a significant interval increase of diffuse craniospinal leptomeningeal metastases of myxopapillary ependymoma and CSF enhancement in the basal cisterns (Fig 5). The enhancing infundibular lesion also was larger.

Discussion

Ependymomas are primary central nervous system neoplasms composed of, and usually derived from, differentiated ependymal cells. Ependymomas constitute approximately 8% of all intracranial gliomas in children and 63% of primary intraspinal intramedullary gliomas (4). Ependymomas occur most often in the posterior fossa in children, arising in the fourth ventricle, followed by the lateral ventricles, third ventricle, aqueduct of Sylvius, spinal cord, cauda equina, and extraventricular hemispheres (7). Pathologically, there are four subtypes of ependymomas: typical ependymoma, malignant ependymoma, subependymoma, and myxopapillary ependymoma (4, 7). The mucinous change of the connective tissue stroma is characteristic of the myxopapillary ependymoma.

The peak prevalence of myxopapillary ependymomas of the spinal cord is in persons 30 to 40 years old (mean age, 36.4 years); 19% of cases present in the first two decades (6). There is a male predominance with a male-to-female ratio of 1.7:1. Spinal myxopapillary ependymomas represent 27% of all spinal ependymomas and occur almost exclusively (95%) in the filum terminale and cauda equina. Cervicothoracic spinal tumors have been reported (6). Intracranial myxopapillary ependymoma is extremely rare. Only two cases, one in a lateral ventricle and one in the cerebral parenchyma of the occipital lobe, have been reported (8, 9).

In our case, the multiple intradural extramedullary nodular enhancing lesions involving the nerve roots of the cauda equina and in the distal thecal sac suggested drop central nervous system metastases from intracranial tumors such as glioma, medulloblastomas, and ependymomas or nonneurogenic neoplasms like lymphoma and leukemia (10). These lesions also could represent the primary site of ependymomas, neurofibromas, schwannomas, or paragangliomas. Other differential diagnostic considerations included tuberculosis, sarcoidosis, and fungal infection (10). The radiographic differential diagnosis for a mass lesion in the in-

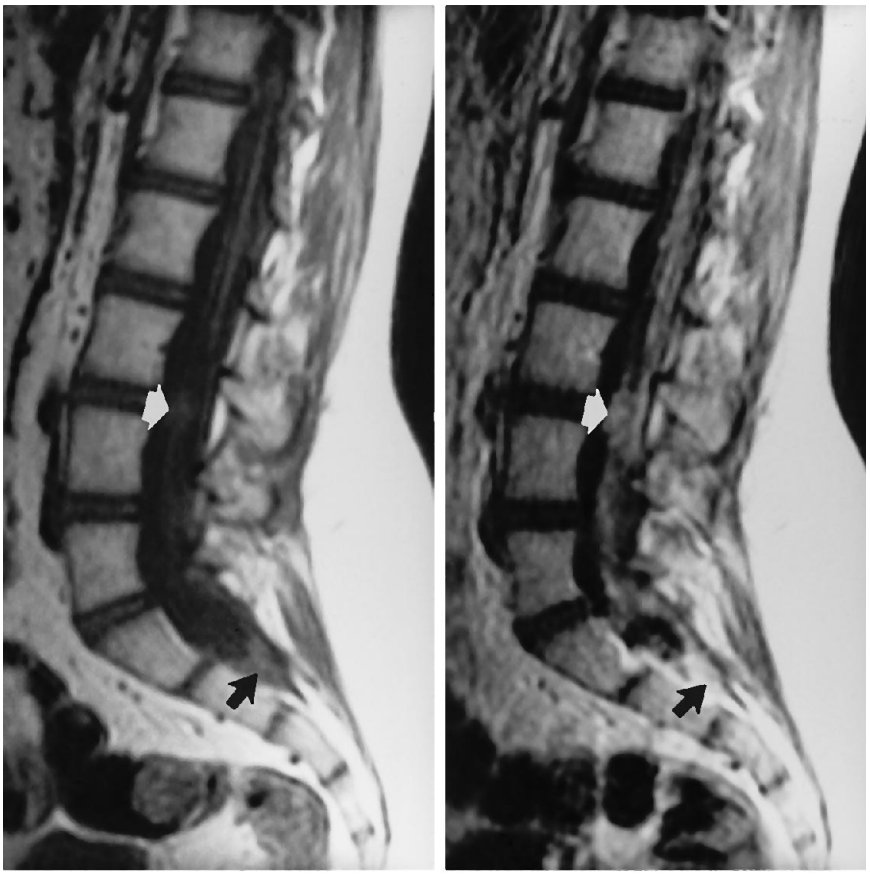

Fig 2. Preenhancement (*A*) and postenhancement (*B*) sagittal T1-weighted spin-echo images (500/11) of the lumbosacral spine. Note the heterogeneously enhanced nodules on the cauda equina (*white arrows*) and at the terminal thecal sac (*black arrows*).

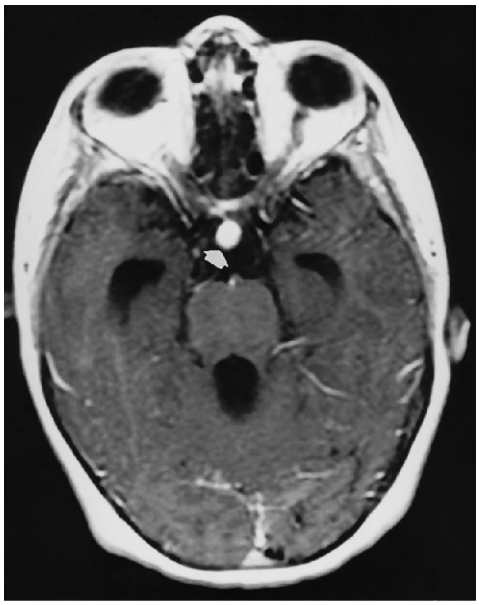

Fig 3. Contrast-enhanced axial T1-weighted spin-echo image (366/11) before treatment shows a markedly enhanced and thickened pituitary infundibulum (*arrow*) without cranial leptomeningeal enhancement.

fundibulum and hypothalamus in the pediatric age group is quite narrow and should include hypothalamic glioma, craniopharyngioma, Langerhans cell histiocytosis, germinoma, meningitis, lymphoma, leukemia, and metastasis (11).

Although the site of origin remains unknown in our case, we suggest that the diffuse leptomeningeal myxopapillary ependymomatosis of the spinal cord probably originates in the region of the cauda equina and spreads through the CSF retrogradely to the region of pituitary infundibulum and antegradely to the distal thecal sac. Retrograde spreading of ependymoma from the spinal cord to the ventricular system is extremely rare and occurs much less often than in the opposite direction (12). The retrograde mechanism seems more likely because of the following facts. First, histologically, 95% of myxopapillary ependymomas occur in the filum terminale and cauda equina (6). Second, reports of retrograde spreading of myxopapillary

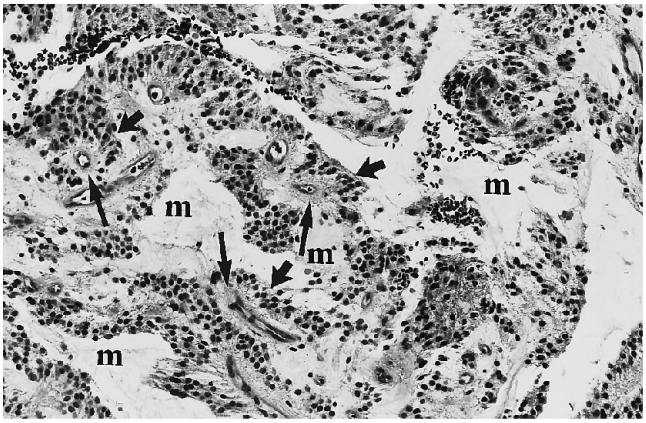

Fig 4. Microscopy of the biopsy specimen of the tumor coating the cauda equina. High-power photomicrograph (hematoxylin-eosin stain, ×40 objective) of myxopapillary ependymoma shows fibrillated cells (*short arrows*) surrounding numerous small vessels with perivascular anuclear zones ("pseudorosettes") (*long arrows*). The fibrillated cells exhibit a blend of glial and epithelial features. There is mild to moderate mucin (*m*) accumulation between islands of tumor cells.

ependymomas have been published (3, 5). Davis and Barnard (3) reported three patients who had lumbosacral myxopapillary ependymomas that spread retrogradely to the left frontoparietal lobe in one patient and to the right cerebellar hemisphere in the other two. In a series of seven pediatric cases of myxopapillary ependymoma of the filum terminale and cauda equina, one patient had metastases to the infundibular part of the third ventricle, hypothalamus, suprasellar region, and left lateral ventricle and another had only suprasellar metastases (5). Surgical excision of the primary lesion occurred before intracranial seeding in all of these five cases, verifying that it is possible for a tumor to have a spinal origin with a solitary intracranial or suprasellar metastasis without significant cranial leptomeningeal spread.

Although less likely, it is possible that the pituitary infundibulum could be the primary site of the myxopapillary ependymoma and the spinal lesions represented drop central nervous system metastases. Primary intracranial myxopapillary ependymoma is very rare, with only two such cases reported. On the other hand, in support of a primary intracranial origin is the fact that definable intraspinal primary lesion was not found at surgery. Characteristically, myxopapillary ependymomas of the cauda equina or the filum terminale grossly appear as well-defined encapsulated, oval or lobulated, soft or firm masses measuring several centimeters (2–6). In our case, no solitary or encapsulated mass was identified at the cauda equina or filum terminale. This appearance is atypical for

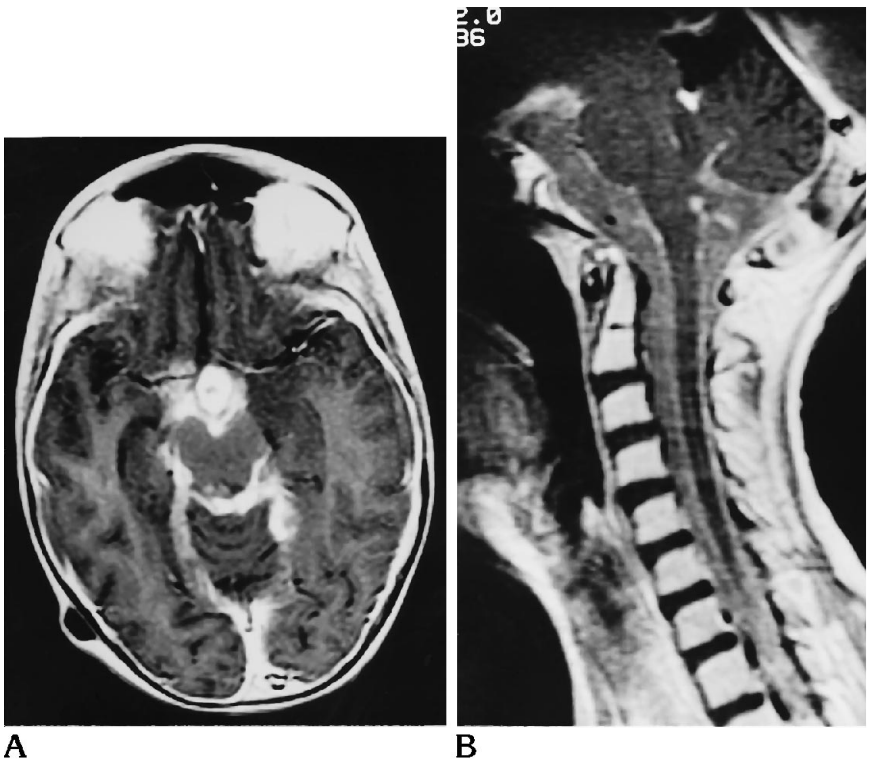

Fig 5. Axial (366/11, *A*) and sagittal (350/19, *B*) contrast-enhanced T1-weighted spin-echo images at 6-month follow-up after chemotherapy and radiation treatment show significant interval increase of diffuse craniospinal leptomeningeal metastases of myxopapillary ependymoma and CSF enhancement of basal cisterns. The enhancing infundibular lesion is also larger. There is no mass lesion or change of the enhancement in the fourth ventricle.

myxopapillary ependymomas arising from the cord. However, this tumor might not be encapsulated and can be invasive in the subarachnoid space and impossible to remove totally (2, 6).

A definite site of origin is impossible to determine in this case because both the infundibular lesion and diffuse spinal leptomeningeal myxopapillary ependymomatosis were shown on the initial MR study. The possible occurrence of synchronous tumors is statistically unlikely but cannot be completely ruled out without biopsy of the infundibular lesion.

Tumor recurrence and metastases are more likely to develop when subtotal resection is the primary mode of treatment, compared with complete excision and irradiation after subtotal resection. The 5-year survival rate for myxopapillary ependymoma is reported as 85% to 100% (5). Six-month follow-up MR in our patient showed increased craniospinal leptomeningeal metastases and CSF enhancement of basal cisterns and fourth ventricle. Failure to respond to chemotherapy and radiation treatment suggested a poor prognosis once diffuse ependymomatosis had occurred.

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