Intraspinal familial clear cell meningioma in a mother and child

Case report

JASON A. HETH, M.D, PATRICIA KIRBY, M.D., AND ARNOLD H. MENEZES, M.D.

Division of Neurosurgery, and Department of Pathology, University of Iowa Hospitals and Clinics, Iowa City, Iowa

The authors present a case of familial clear cell meningioma in which the proband is a child with an intraspinal tumor. The clear cell meningioma variant has recently been studied. The literature regarding clear cell meningioma is reviewed.

KEY WORDS • meningioma • clear cell • familial tumor • children

MENINGIOMAS are relatively common intracranial and spinal tumors in adults but their occurrence in children is uncommon. Many case reports have documented familial meningioma and current debate centers on whether familial meningiomas are allelic to neurofibromatosis. Clear cell meningioma is a variant that has only recently been described. We present the cases of a 7-year-old girl and her mother who each harbored a familial clear cell meningioma.

Case Reports

Case 1

This 7-year-old right-handed girl presented with a 4-month history of lower back pain that radiated to her right leg. Forward bending caused her significant back pain.

Examination. Physical examination revealed neither café-au-lait spots nor neurofibromas. Her cranial nerve function was normal. She had full motor function but had hypesthesia and hypalgesia on the sole of her left foot. Her lower-extremity reflexes were absent, and she had positive straight-leg raising signs bilaterally. Magnetic resonance imaging of the lumbar spine demonstrated a smooth-bordered, uniformly enhancing, intradural extramedullary mass occupying the L4–5 spinal level (Fig. 1).

Operation and Postoperative Course. The patient underwent L3–5 osteoplastic laminectomies, at which time a 26 × 14 × 15–mm mass was resected en bloc. The pathological diagnosis was clear cell meningioma (Fig. 2). Postoperatively, she demonstrated no new deficits, and she remains recurrence free at 13 months.

Case 2

This 31-year-old right-handed woman, the mother of the child reported in Case 1, underwent a posterior fossa craniectomy with subtotal resection of a foramen magnum clear cell meningioma 4 months prior to presentation at our institution. Two months later, she developed recurrent nausea, diplopia, dizziness, and epigastric distress.

Examination. On physical examination, she was awake and alert. Cranial nerve examination was remarkable for mild dysfunction of her left fifth and sixth cranial nerves as well as diminished left-sided gag reflex, a hypomobile left palate, and left-sided tongue atrophy. She had full motor strength, but sensory examination revealed right-sided hypesthesia and hypalgesia. Deep tendon reflexes were 2+. She exhibited dysdiadochokinesia of her left arm and leg. She did not harbor café-au-lait spots or cutaneous neurofibromas. Serial imaging MR imaging of the brain demonstrated an enhancing mass at the left anterolateral aspect of the foramen magnum, causing brainstem displacement to the right (Fig. 3). Angiography revealed no significant tumor vascularity.

Operation and Postoperative Course. The patient underwent a far-lateral transcondylar approach for gross-total tumor resection. Pathological examination revealed a clear cell meningioma (Fig. 4). Postoperatively, she sustained only a mild increase in ataxia and her other deficits remained stable. She remains free of recurrence at 3 years.

Abbreviations used in this paper: MR = magnetic resonance; NF = neurofibromatosis; VHL = von Hippel–Lindau.
Family History

The mother in our Case 2 has a maternal stepsister who has two daughters who have had meningiomas (Fig. 5). The oldest daughter had a posterior fossa meningioma and the second oldest daughter had a lumbar spine meningioma. No family member, including these two females, has any stigmata of NF.

Discussion

We present an interesting case of a familial clear cell meningioma in which the proband is a child with an intraspinal meningioma. The clear cell meningioma variant has only recently been described and studied. Zorluemir, et al. reviewed 13 cases of clear cell meningioma (seven female and six male patients). Six of these patients were seen at the Mayo Clinic and they represented 0.2% of the Mayo’s experience with meningioma. Seven of the 13 tumors were intraspinal (five lumbar, one lumbosacral, and one thoracic). The lesions were described as being smooth and bosselated and appearing solid gray-pink to tan-red. Microscopically, the clear cell meningiomas were moderately cellular with polygonal cells having prominent intracytoplasmic glycogen. Their cases included two children with an intraspinal tumor. Recurrence occurred in 61% of the cases they reported with cranial to spinal metastasis in 8%. Twenty three percent of their patients died as a result of meningioma. Dubois and colleagues reported on a 10-year-old girl with a clear cell meningioma of the cauda equina. Their patient is similar to our patient (Case 1) in that pain was a primary presenting complaint; the MR imaging characteristics in these cases are also similar. There are also two reports of lumbar and sacrococcygeal clear cell meningiomas that occurred in 32- and 38-year-old men, respectively. The natural history of meningioma growth favors discovery well out of the pediatric age range. Periulong, et al. reviewed the cases of 20 children with meningiomas who were treated at the Childrens Hospital of Philadelphia; these cases represented 2% of pediatric brain tumors diagnosed between 1975 and 1991. Four of these tumors were intraspinal (0.4% spinal meningioma). Sheikh, et al. reported a meningioma incidence of 2.8% in children treated from 1981 to 1993; none of the children harbored a spinal tumor. Mallucci and others reported 21 meningiomas (1.62%) of 1294 pediatric central nervous system tumors identified between 1957 and 1993 (one case was intraspinal). Baumgartner and Sorenson found that 4% of all meningiomas (14 cases) diagnosed from 1964 to 1994 occurred in children. One tumor was in the cervical spine. Shih and colleagues reported on a 12-year-old boy with

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a cerebellopontine angle clear cell meningioma. These studies showed a meningioma incidence of 1.61 to 4% with a median or mean patient age of 10 to 11 years. The case reported by Shih, et al., is the only one identified as a clear cell variant. The characteristics of all clear cell meningiomas reported in the English literature are summarized in Table 1. The literature, however, contains case reports of spinal meningiomas in children. These children presented with pain and paraparesis or quadriplegic paresis and the tumors were found in both epidural and intradural extramedullary locations.

Familial inheritance of meningiomas is well known. These occur primarily in association with NF2 and VHL diseases. Maxwell and colleagues collected 14 cases of isolated familial meningiomas. Our patients do not have the stigmata of either NF or VHL. Stronger proof of a familial relationship has been difficult to establish; almost all of the laboratories we contacted do not perform linkage analysis unless NF has already been diagnosed in one family. One laboratory will perform an analysis; however, reimbursement remains a difficult issue to resolve. Our case highlights the fact that even for relatively common genetic diseases, genetic evaluation may be difficult to obtain.

We report on a child with a familial spinal clear cell meningioma. Neither mother nor child has stigmata or evidence of NF or VHL disease, although these diseases may certainly declare themselves in the future. Currently, these two patients would be considered an isolated case of familial meningioma. The aggressive nature of the mother’s tumor is not unusual. The patient required two resections within 5 months to control her tumor. However both child and mother are without recurrence at 1 and 3 years, respectively. Zorludemir, et al., believed that clear cell meningiomas were a more aggressive subtype. The demographics shown in Table 1 support this characterization in the pediatric population, as 60% of the clear cell meningioma...
<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Cases</th>
<th>Sex, Age (yrs)</th>
<th>Location</th>
<th>Surgery</th>
<th>Follow-Up Results</th>
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<tr>
<td>Shiraishi, 1991</td>
<td>1</td>
<td>M, 32</td>
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<td>Shih, et al., 1996</td>
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<td>M, 12</td>
<td>CPA</td>
<td>GTR</td>
<td>no recurrence at 5 mos</td>
</tr>
<tr>
<td>Dubois, et al., 1998</td>
<td>1</td>
<td>F, 10</td>
<td>L1–4</td>
<td>GTR</td>
<td>recurrence 6 mos</td>
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<tr>
<td>Imlay, et al., 1998</td>
<td>1</td>
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<td>posterior fossa</td>
<td>STR</td>
<td>metastatic disease in sacrum 7 yrs postop</td>
</tr>
<tr>
<td>Matsui, et al., 1998</td>
<td>1</td>
<td>F, 9</td>
<td>cauda equina</td>
<td>STR</td>
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<td>Maxwell, et al., 1998</td>
<td>2</td>
<td>M, 24</td>
<td>L-5</td>
<td>GTR</td>
<td>no recurrence (+M)</td>
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<td>Pimentel, et al., 1998</td>
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<td>F, 24</td>
<td>tentorium–clinoid</td>
<td>STR</td>
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<td>Holtzman &amp; Jormark, 1996</td>
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<td>GTR</td>
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<td>Dubois, et al., 1998</td>
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<td>cervical</td>
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<td>no recurrence 45 mos</td>
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<td>Teo, et al., 1998</td>
<td>1</td>
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<td>intraparenchymal brainstem</td>
<td>STR</td>
<td>?</td>
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<td>M, 42</td>
<td>lumbar sacral cord</td>
<td>GTR</td>
<td>no recurrence at 24 mos</td>
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<td>Lee, et al., 2000</td>
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<td>parietal lobe</td>
<td>GTR</td>
<td>recurrence at 7 mos</td>
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<tr>
<td>present report</td>
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<td>F, 7</td>
<td>L4–5</td>
<td>GTR</td>
<td>no recurrence (+f)</td>
</tr>
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</table>

* CPA = cerebellopontine angle; DOD = died of disease; +f = positive family history; GTR = gross-total resection; +M = positive Merlin staining; STR = subtotal resection.
Intraspinal familial clear-cell meningioma

giomas recurred. The age of the patient in Case 2 at presentation is unusual, as the location of her tumor. Meningiomas in the pediatric population are uncommon and spinal meningiomas in this population are presented as case reports.\textsuperscript{2,6} Table 1 does show, however, that the proportion of spinal to total meningiomas is higher for clear cell meningioma (45\%) than for all total meningiomas (the highest being 20\% of the following reported cases\textsuperscript{2,12,15,18}).

The differential diagnosis for a lumbar intradural mass includes schwannoma, neurofibroma, ependymoma, drop metastasis from a medulloblastoma, neuroblastoma, or pineoblastoma. Case 1 shows that meningioma should be considered in this differential diagnosis as well.

The operative goal with the clear cell meningioma is gross total en bloc resection. En bloc resection is favored to prevent leaving microscopic nests of cells of this meningioma behind as a nidus for recurrence. As shown in Table 1, although 47\% of the patients in whom gross-total resections were performed suffered recurrence, 86\% of those in whom subtotal resections were performed and who survived the immediate postoperative period suffered recurrence.

Conclusions

Clear cell meningioma is a recently described variant of meningioma. It is an important variant because of its aggressive behavior and recurrence potential. We describe a clear cell meningioma of the lumbar spine in a girl whose mother also harbored a clear cell meningioma. Clear cell meningioma may be familial and this report reinforces the rarity of spinal meningiomas in the pediatric population as well as the recurrence potential of the clear cell meningioma.

References


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Address reprint requests to: Arnold Menezes, M.D., Division of Neurosurgery, University of Iowa Hospitals and Clinics, 200 Hawkins Drive, Iowa City, Iowa 52242.