A Case of Cutaneous Meningioma in the Rudimentary Meningocele

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ABSTRACT

We report a rare case of neonatal cutaneous meningioma derived from a rudimentary meningocele. This neonate had a congenital skin-covered hump in the thoracolumbar region. Computed tomography showed bifid laminae of T12 and L1 underneath the mass lesion. Magnetic resonance images showed the mass to have no cerebrospinal fluid space and that it had a stalk connecting to the spinal canal. Split cord malformation was also observed under the bifid laminae. Because of the increasing size of the lump and cosmetic reasons, the parents opted for surgical treatment. We operated on the patient 9 months after birth. Operative findings showed that the cutaneous mass was connected to intraspinal contents by a vascular stalk and it was totally removed. The split spinal cord was untouched. The histopathological findings of the mass showed components of meningioma with a collagenous matrix. We concluded that this patient had a meningioma derived from rudimentary meningocele.

Key words: Cutaneous meningioma, Rudimentary meningocele, Spine

Rudimentary meningocele is a rare congenital anomaly found along the midline in the head and back of neonates. It is considered to be an abortive type of meningocele as the skin mass has only a cluster of meningeal cells without space for the cerebrospinal fluid. In some instances, components of meningioma are found in the skin mass of rudimentary meningoceles. It is unclear whether or not this meningioma behaves like a growing neoplasm. We report a case of cutaneous meningioma in the rudimentary meningocele with discussions of nomenclature and neoplastic significance of this clinical entity.

CASE REPORT

This neonate presented with a congenital mass lesion in the thoracolumbar region. As the mass was covered by the skin and he had no apparent neurological deficit, the parents opted for a wait and see policy. Neither developmental disorder nor neurological deficit was evident thereafter.

Nine months after birth, the parents wished for surgical removal of the cutaneous mass because of a gradual increase in size of the mass and for cosmetic reasons (Fig. 1). Both T1- and T2-weighted sagittal magnetic resonance (MR) images revealed a low signal intensity mass lesion at L1 level. There was a stalk extending from the mass to the spinal canal and split cord malformation at T12-L1 level. However, the relationship between the mass lesion and the anomalous spinal cord was unclear (Fig. 2, A-C). Computed tomography showed bifid laminae of T12 and L1 beneath the mass lesion (Fig. 2D). The patient was placed in a prone posi-

Fig. 1. Appearance of the cutaneous nodule in the thoracolumbar region.
resected en-bloc after resecting the artery (Fig. 3). Intradural inspection revealed a split in a region of spinal cord without bony or dural septa which was left untouched.

The mass was composed of fibro-collagenous tissue with a cluster of meningothelial cells. Proliferated meningothelial cells were arranged in a whorl pattern with positive staining for epithelial membrane antigen. These findings indicated the histological character of meningioma (Fig. 4).

**DISCUSSION**

**Cutaneous meningioma and rudimentary meningocele**

Lopez et al classified cutaneous meningiomas into 3 types based on their etiologies. Type I is a congenital type, mostly found on the scalp and paravertebral regions, in which meningioma arise from ectopic meningothelial cells trapped in the dermis through developmental defects such as the failure of neural tube closure. Type II is ectopic meningioma found around the eyes, ears, nose and mouth and derived from the remnant arachnoid cells extending along cranial nerves. Type III is a meningioma extending to the skin directly from the intracranial or intraspinal primary lesions. The present case had a component of meningioma in the cutaneous mass of the thoracolumbar region. The mass had a vascular stalk connecting to the intraspinal content through the bifid laminae. The findings of the present case correspond with those seen in type I cutaneous meningioma.

Rudimentary meningoceles are a developmental anomaly characterized by a mass in the posterior midline along the cranio-spinal axis. According to Stone’s review of 47 cases of rudimentary meningocele, 37 lesions were located on the head,
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and rudimentary meningocele have been proposed: hamartoma of the scalp, sequestered meningocele, acoelic meningeal hamartoma, and cutaneous heterotopic meningeal nodules\(^2\). Strictly speaking, type I cutaneous meningioma is not a synonym for rudimentary meningocele because rudimentary meningoceles do not always have meningioma components. We, therefore, prefer to classify this case as a cutaneous meningioma in rudimentary meningocele.

With regard to meningiomas in spinal rudimentary meningoceles, we obtained 4 such cases including our own (Table 1)\(^3,7,8\). Interestingly, 3 out of 4 cases underwent surgery after 10 years old even though a cutaneous mass had been noticed on their backs much earlier. All cases had bifid laminae beneath the cutaneous mass. Three cases had no neurological deficit and one was complicated by hemiparesis and hemihypoesthesia, probably due to scoliosis. Surgical findings disclosed that 2 cutaneous masses had attachments to the dura and 2 had fibrous attachments to the spinal cord. All surgeries were performed without additional neurological sequelae.

<table>
<thead>
<tr>
<th>Author</th>
<th>age</th>
<th>sex</th>
<th>spinal level</th>
<th>bifid lamina</th>
<th>neurological deficit</th>
<th>attachment of the cutaneous mass</th>
<th>histopathological findings</th>
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<tbody>
<tr>
<td>Zaaroo(^8)</td>
<td>13</td>
<td>F</td>
<td>lumbar</td>
<td>L4</td>
<td>none</td>
<td>dura</td>
<td>meningeal whorl formation, psammoma bodies, collagen bundles</td>
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<tr>
<td>Kaplan(^7)</td>
<td>11</td>
<td>F</td>
<td>cervical</td>
<td>C3-C4</td>
<td>hemiparesis</td>
<td>dura</td>
<td>psammoma bodies, collagen bundles</td>
</tr>
<tr>
<td>Wang(^7)</td>
<td>21</td>
<td>F</td>
<td>cervical</td>
<td>C3</td>
<td>none</td>
<td>dura and spinal cord</td>
<td>meningeal whorl formation, psammoma bodies, collagen bundles</td>
</tr>
<tr>
<td>Present case</td>
<td>0</td>
<td>M</td>
<td>thoraco-lumbar</td>
<td>T12-L1</td>
<td>none</td>
<td>dura and spinal cord</td>
<td>meningeal whorl formation, psammoma bodies, collagen bundles</td>
</tr>
</tbody>
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Fig. 4. Photomicrographs of the resected skin mass
A: Multiple nests of round and oval meningothelial cells with basophilic nuclei and dispersed chromatin, alongside psammoma bodies in fibrocollagenous matrix (HE, \(\times 200\)).
B: Meningothelial cells revealed cytoplasmic and membrane positivity for epithelial membrane antigen (EMA, \(\times 200\)).
Meningioma in the rudimentary meningocele: its significance as a viable tumor

It has not been elucidated whether meningioma in the rudimentary meningocele actually behave as a viable and growing neoplasm. Since most of these lesions are resected early after the birth, a long-term history of meningioma in the cutaneous mass has rarely been reported. Brantsch et al reported a 33-year-old female presenting with a scalp nodule existing since birth. Her nodule exhibited painful growth after the delivery of the second child. Surgical exploration of the nodule showed subcutaneous meningioma with no evident connection to the intracranial contents. The authors concluded meningioma might have developed from meningothelial cells in a rudimentary meningocele. Wang et al reported a 21-year-old woman presenting with a gradually growing mass in the posterior cervical region. Preoperative neuroimaging studies showed a skin mass connecting to the intraspinal contents through the bifid C3 lamina. She underwent resection of the mass for cosmetic reasons. Histopathological findings of the resected mass showed clusters of meningocytes and a psammoma body. They concluded that this case was cutaneous meningioma growing in the rudimentary meningocele. These two reports may suggest that untreated meningioma in the rudimentary meningocele may grow and act as a true neoplasm. Early resection of the rudimentary meningocele might be justified even if it causes no neurological deficit.

CONCLUSION

The authors report a case of cutaneous meningioma in the rudimentary meningocele. Although the natural history of this disease is unclear, early surgical intervention seems to be justified because of its possible neoplastic nature and for cosmetic reasons.

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