Rudimentary Meningocele of the Scalp

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Rudimentary meningocele, a variant of primary cutaneous meningioma, is a rare embryologic malformation with meningeal tissue occurring in the skin. This work studied five cases of this condition. All five cases were congenital and presented as alopecic lesions on the occipital area. No underlying bony defect or communication to the intracranial compartment was detected by brain computed tomography (CT) or echoencephalogram. Histologically, the major changes were mostly concentrated in the subcutis, where strands of meningotheelial cells were embedded in dense collagenous tissue. The meningotheelial cells were immunopositive for vimentin and epithelial membrane antigen and lacked cytokeratin, S-100 protein, smooth muscle actin and endothelial markers. The related literature also was reviewed. (Dermatol Sinica 21 : 394-401, 2003)

Key words: Epithelial membrane antigen (EMA), Primary cutaneous meningioma, Rudimentary meningocele, Vimentin

INTRODUCTION

In 1974, Lopez et al. reviewed 25 cases of cutaneous meningioma and classified them into three groups.¹ Type I or primary cutaneous meningioma (PCM) occurs on the scalp, face, or paravertebral regions of children and young adults. No intracranial or intravertebral tumor is involved. This type is usually present at birth
and follows a benign course. Type II corresponds to ectopic meningioma arising from arachnoid cells that accompany the cranial and spinal nerves. This type generally appears in adults as a de novo lesion and represents a cutaneous extension from an ectopic soft tissue meningioma. Finally, type III represents direct extension into the skin from an intracranial meningioma through a foramen or a bone defect. This study reports five cases of rudimentary meningocele, a variant of PCM, examined via histopathology, immunohistochemistry, and imaging studies.

**Patients and Methods**

All cases labeled meningocele were retrieved from the surgical pathology file of the Department of Pathology, Chang Gung Memorial Hospital, Taiwan. Cases involving the central nervous system were excluded. This approach yielded a total of five during 2000 to 2001. All clinical data, including age, sex, clinical appearance and symptoms, were recorded. Formalin-fixed and paraffin-embedded tissue from these five skin lesions was examined by histopathology and immunohistochemical study. The immunohistochemical stains applied to the five cases included monoclonal antibodies directed against epithelial membrane antigen (EMA) (Dako, 1:50), vimentin (Dako, 1:200), and polyclonal antibodies directed against S-100 protein (Dako, 1:1000). Two cases were further assessed with anti-CD34 (Dako, monoclonal, 1:50); one of these cases was also assessed with anti-cytokeratin AE1/AE3 (Biogenex, monoclonal, 1:200) while the other was also assessed with anti-smooth muscle actin.

**Fig. 1**

Clinical pictures. (a) A congenital, flesh-colored, alopecic, soft nodule occurred on the paramedian occiput of Case 3.

(b) Grouped bluish alopecic patches with central atrophy located on the midline occiput of Case 2.
RESULTS

CLINICAL INFORMATION

Table I summarizes the clinical data. The five patients enrolled in the study were children ranging in age from 3 days to 11 years. All lesions were congenital and located on the midline or paramedian occipital scalp. The lesion sizes ranged from 1 to 4.5 cm. All lesions but one were solitary. The lesions were round or oval and presented as flesh-colored or erythematous patches, plaques or nodules (Fig. 1a).

The skin lesion of Patient 2 comprised five atrophied patches (Fig. 1b). All lesions were alopecic, and no one exhibited a “hair collar sign”. No symptoms such as pain, tenderness, or itching were noted. The skin lesions of Patients 1 and 2 slowly grew with the child, but growth stopped around 6 years of age. No family history existed except in Patient 2, whose sister also had a similar lesion over the midline occiput since birth. Brain CT or echoencephalogram displayed no intracranial lesion or bony

Table I. Clinical Features

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at diagnosis/sex*</th>
<th>Age of onset</th>
<th>Clinical findings</th>
<th>Size, cm/Site</th>
<th>Other anomalies</th>
<th>Imaging studies</th>
<th>Treatment/Follow-up period</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11yo/F</td>
<td>Birth</td>
<td>An alopecic erythematous infiltrated plaque</td>
<td>4.5X4/Right paramedian occiput</td>
<td>None</td>
<td>Brain CT: no bone defect or intracranial lesion</td>
<td>None/ No enlargement after 6yo</td>
</tr>
<tr>
<td>2</td>
<td>5yo/F</td>
<td>Birth</td>
<td>Grouped bluish to grayish-red alopecic patches</td>
<td>3.5X3/Midline occiput</td>
<td>None</td>
<td>Brain echogram (in infant): normal</td>
<td>None/ No enlargement at 7yo</td>
</tr>
<tr>
<td>3</td>
<td>8mo/F</td>
<td>Birth</td>
<td>An alopecic flesh-colored nodule</td>
<td>1.2X1/Paramedian occiput</td>
<td>None</td>
<td>Brain echogram: normal</td>
<td>None/ no obvious enlargement at 1.5yo Total excision/ no recurrence at 2.5yo</td>
</tr>
<tr>
<td>4</td>
<td>15mo/M</td>
<td>Birth</td>
<td>An alopecic flesh-colored subcutaneous tumor with overlying atrophied skin</td>
<td>4X4/Right paramedian occiput</td>
<td>None</td>
<td>Brain CT: normal</td>
<td>Total excision/ no recurrence at 2 yo</td>
</tr>
<tr>
<td>5</td>
<td>3do/M</td>
<td>Birth</td>
<td>An alopecic flesh-colored nodule</td>
<td>1X1/Midline occiput</td>
<td>None</td>
<td>Brain echogram: normal</td>
<td>Total excision/ no recurrence at 2 yo</td>
</tr>
</tbody>
</table>

*F: female; M: male
CT: computed tomography
yo: year old
mo: month old
do: day old
defects (Fig. 2). No cerebrospinal fluid (CSF) leakage occurred during surgery. Two patients (Patients 4 and 5) received complete excision of the mass, with no recurrence during 2.5 years of follow up. Meanwhile, Patients 1 and 2 refused further surgical management after skin biopsy because of a lack of life quality impacting symptoms and because the mass had stopped enlarging. Finally, Patient 3 decided to delay surgery owing to the absence of any noticeable tumor growth.

HISTOPATHOLOGIC FINDINGS

The five lesions displayed a similar histologic appearance. They were located in the subcutaneous tissues with/without extension into the deep dermis. The meningothelial cells were recognizable by their ovoid or spindle shape with scant eosinophilic cytoplasm and pale, round, and vesicular nuclei (Fig. 3 & 4). The cells formed strands, small nests or thin-walled pseudovascular spaces and encompassed the surrounding collagen fibers and adnexal structures (Fig. 3 & 4). Cystic structures lined by meningothelial cells with papillary projections were noted in cases 3 and 5 (Fig. 4). Moreover, the meningothelial cells aggregated into whorls in case 1, 4 (Fig. 5a). Scattered bipolar melanocytes were also observed in case 2. The hair follicles of these skin specimens were atrophied or reduced in number. The number of other adnexal structures (apocrine or eccrine sweat glands) also changed in some cases. No calcification or psammoma body was noted in any of the five cases.

IMMUNOHISTOCHEMISTRY

Immunohistochemical studies showed diffuse reactivity for vimentin (VIM) and epithe-
Epithelial membrane antigen (EMA) in all lesional cells (Fig. 5c-5d). All lesions stained negative for S-100 protein. Other immunohistochemical studies, such as CD34, cytokeratin AE1/AE3, or smooth muscle actin, revealed negative results where performed. Table II summarizes the immunohistochemical studies for the five subject cases.

**DISCUSSION**

Rudimentary meningocele is a rare developmental anomaly in which meningotheial elements are displaced into the skin and subcutaneous tissue. Previously, rudimentary meningocele has been described under various different terms, including cutaneous meningioma, hamartoma of the scalp with ectopic meningotheial elements, and meningotheilomatous hamartoma. To date, only around 50 cases have been reported worldwide. This investigation is the first to analyze this entity in Taiwan.

Rudimentary meningocele occurs at birth. Clinically, this condition frequently occurs on the scalp (72%), especially the parietal and occipital areas, and on the midline and paravertebral regions of the back. The lesions are slow-growing, well-circumscribed, and generally asymptomatic. The lesions present as patches, papules, nodules, or exophytic masses. The lesions are occasionally alopecic, and rarely are hypertrichotic with or without congenital melanocytic nevus on the lumbar region. A hair collar sign, a ring of dark hair encircling the lesion, also may occur. Clinical differential diagnoses include aplasia cutis congenita, sebaceous nevus, lipoma, epidermal inclusion cyst, adnexal tumor, lymphangioma, alopecia areata, fibroma, skin tag, congenital melanocytic nevus and dermoid cyst. Aplasia cutis congenita is characterized by a congenital, circumscribed area of alopecia and scarring on the scalp. The presence of hair collar sign should alert practitioners to the possibility of a significant defect, such as meningocele. The five subject cases of rudimentary meningocele had been present since birth, but had different presentations, including patch (or patches), plaque or nodule, with or without skin discoloration. Incidentally, all five lesions were located on the midline or paravertebral area of the occiput. Moreover, all of the lesions were asymptomatic and alopecic. No hair collar sign was noticed. Generally, no family history existed, though rudimentary memingocele seemed to occur in both siblings of our case 2. Miyamoto et al. also had reported two siblings of this condition.

**Fig. 5a**
Spindle and satellite meningotheial cells were dispersed among the coarse collagen bundles and arranged into a whorl. (Hematoxylin-eosin stain, x400)

**Fig. 2**
The meningotheial cells displayed pale-staining cytoplasm, round or oval vesicular nuclei, and small and bland nucleoli. Collagen bodies and multinucleated syncytial-like giant cells were also present. (Hematoxylin-eosin stain, x600)
But for such a rare condition, it does seem like rather a large coincidence.

The microscopic features are often subtle, with meningeal tissue simulating the appearance of vascular or connective tissue. Typically, rudimentary meningocele is situated in the subcutaneous tissue with variable extension into the dermis. Moreover, there typically is no involvement of the superficial corium or communication with the epidermis. The lesions contain a myxoid stroma partitioned by numerous thin-walled pseudovascular spaces lined by oval to spindle-shaped meningothelial cells. These “channels” are separated by variably dense collagen fibers and are devoid of erythrocytes. The meningothelial cells are characteristically uniform and have well-defined, oval or rounded basophilic nuclei with dispersed, finely stippled chromatin and faintly eosinophilic cytoplasm. The proliferating meningothelial cells are usually aggregated into abortive whorls or tactoids. Moreover, the proliferating cells may lack a distinct cell membrane with HE stain, leading to the appearance of syncytial giant cells. Psammoma and collagen bodies may be present around the cellular area. Furthermore, decreases or increases in adnexal structures may also occur. The results of this study were comparable to previously published reports, but no detectable calcification or psammoma body was noted in our cases.

Immunohistochemical studies revealed diffuse reactivity for vimentin in virtually all lesional cells. The cells are usually labeled by anti-epithelial membrane antigen (EMA). Antibodies directed against S-100 protein, keratin, smooth muscle actin, desmin, neuron specific enolase, GFAP (glial fibrillary acidic protein) failed to label meningothelial cells. However, the multinucleated dendritic elements in the lesion may be stained variably for S-100 protein. Similar immunohistochemical findings also were observed in the subject cases (Table II).

Rudimentary meningocele (RM) previously has been regarded by some authors as a form of primary cutaneous meningioma. However, such a classification is inappropriate, since it implies that the lesion is a neoplasm rather than a malformation. Sibley and Cooper termed these lesions rudimentary meningoceles (RM), suggesting that they are developmental malformations closely related to classic meningocele (CM). Indeed, RM and CM share similar histopathological and biological features. Specifically, these lesions are distinguishable.

**Fig. 5c**
The meningothelial cells were positively stained for epithelial membrane antigen (EMA). (x400)

**Fig. 5d**
The meningothelial cells were positively stained for vimentin. (x400)
only based on attendant clinical data. RM is relatively localized to the scalp or neck, comes to clinical attention at a later age, and, unlike CM, tends not to be associated with major structural defects in the coverings of the central nervous system. Nonetheless, minor abnormalities of the skull and partial alopecia may occur in association with RM. To exclude the possibility of a connection to the central nervous system, preoperative neurosurgical studies and imaging studies are indicated. Major skeletal or neurologic defects should not occur in RM, which is an indolent growth that is curable by simple excision. The five patients presented here have no skull defects or intracranial lesions. No other congenital anomalies were associated with the subject lesions. Two of the five scalp lesions were totally excised, and no signs of recurrence being noted for more than 2 years. The three patients who did not receive further surgery also experienced no discomfort during follow up. No obvious enlargement or other changes were noted in the skin lesions.

The pseudovascular spaces observed in RM may mimic cutaneous angiosarcoma histopathologically. Lack of immunoreactivity for endothelial markers such as factor VIII-related antigen, CD31, CD34 in RM can help to differentiate RM from vascular tumors.

Table III lists the immunohistochemical comparisons of cutaneous rudimentary meningocele and its diagnostic alternatives.

Table II. Immunohistochemical Findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>EMA</th>
<th>VIM</th>
<th>S-100</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>CD34−, Smooth muscle actin−</td>
</tr>
<tr>
<td>2</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>CD34−, cytokeratin AE1/AE3−</td>
</tr>
<tr>
<td>3</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>Not done</td>
</tr>
<tr>
<td>4</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>Not done</td>
</tr>
<tr>
<td>5</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>Not done</td>
</tr>
</tbody>
</table>

VIM= Vimentin
EMA= Epithelial Membrane Antigen

Table III. Immunohistochemical Comparisons of Cutaneous Rudimentary Meningocele and Other Diseases

<table>
<thead>
<tr>
<th>LESION</th>
<th>VIM</th>
<th>EMA</th>
<th>FVIII</th>
<th>CD 34</th>
<th>UEA</th>
<th>MSA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rudimentary Meningocele</td>
<td>+</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>−</td>
</tr>
<tr>
<td>Classic Meningocele</td>
<td>+</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>−</td>
</tr>
<tr>
<td>Endothelial Neoplasms</td>
<td>+</td>
<td>+</td>
<td>+/−</td>
<td>+/−</td>
<td>+</td>
<td>−</td>
</tr>
<tr>
<td>Giant Cell Fibroblastoma</td>
<td>+</td>
<td>−</td>
<td>+</td>
<td>−</td>
<td>+</td>
<td>+/−</td>
</tr>
</tbody>
</table>

VIM= Vimentin
EMA= Epithelial Membrane Antigen
FVIII= Factor VIII-related antigen
UEA= Ulex europaeus I agglutinin
MSA= Muscle-specific actin
The etiology of rudimentary meningocele is unknown. Some authors consider these lesions to be a form of meningocele with an obliterated intracranial communication, while others classify them as remnants of the neural crest. Another recent theory proposes that multisite initiation of neural tube closure and the subsequent failure of this closure may explain the pathogenesis of rudimentary meningocele.1, 8, 11, 16-18

In summary, rudimentary meningocele is a rare condition involving a developmental malformation closely related to classic meningocele. Generally no major skeletal or neurologic defects are involved. Rudimentary meningocele can be cured by simple excision. The pathogenesis of this condition remains mysterious. However, given the probable pathogenesis from a neural tube defect, imaging studies to exclude any communication to the central nervous system should precede any invasive investigation or intervention. This work demonstrates the different clinical spectrum of this rare disorder and highlights the importance of special immunohistochemical staining in making diagnosis.

REFERENCES