Rudimentary Meningocele: Remnant of a Neural Tube Defect?
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Background: Rudimentary meningocele, a malformation in which meningothelial elements are present in the skin and subcutaneous tissue, has been described in the past under a variety of different terms and has also been referred to as cutaneous meningioma. There has been debate as to whether rudimentary meningocele is an atretic form of meningocele or results from growth of meningial cells displaced along cutaneous nerves.

Objective: We reviewed the clinical, histological, and immunohistochemical characteristics of rudimentary meningocele in an attempt to assess the most likely pathologic mechanism for it.

Design: Retrospective study.

Setting: University hospitals.

Patients: Thirteen children with rudimentary meningocele.

Main Outcome Measures: Medical records were reviewed and histopathologic examination as well as immunohistochemistry studies were performed for each case. A panel of immunoperoxidase reagents (EMA, CD31, CD34, CD57, S-100, and CAM 5.2) was used to assess lineage and to confirm the meningothelial nature of these lesions.

Results: Recent evidence indicating a multisite closure of the neural tube in humans suggests that classic meningocele and rudimentary meningocele are on a continuous spectrum.

Conclusion: Rudimentary meningocele seems to be a remnant of a neural tube defect in which abnormal attachment of the developing neural tube to skin (comparable to that in classic meningocele) could explain the presence of ectopic meningial tissue. In the majority of cases, no underlying bony defect or communication to the meninges could be detected. However, in light of the probable pathogenesis, imaging studies to exclude any communication to the central nervous system should precede any invasive evaluation or intervention.

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RUDIMENTARY meningocele is an uncommon developmental anomaly in which meningothelial elements are displaced into the skin and subcutaneous tissue. In the past it has been described under a variety of different terms, such as cutaneous meningioma, hamartoma (of the scalp) with ectopic meningothelial elements, sequestrated meningocele, acocelic meningial hamartoma, and cutaneous heterotopic meningial nodules.1-8 Controversy exists as to whether this entity represents a form of meningocele in which the underlying connection to the meningial space is obliterated, or whether lesions develop because of proliferation of meningial cells along the routes of cutaneous nerves.9

There is recent evidence for a multisite closure of the neural tube in humans similar to that observed in experimental animals.10,11 Multiple distinct closure sites of the neural tube explain the most frequent locations of neural tube defects. Failure to achieve complete fusion results in various forms of dysraphia.

We present 13 cases of rudimentary meningocele that show a strikingly similar anatomical distribution to the neural tube fusion sites observed experimentally. The multisite closure model explains the congenital nature and distribution of these lesions and also coincides with the distribution of classic meningoceles.

HISTOPATHOLOGIC FINDINGS

The microscopic findings are summarized in Table 2. In general, skin append-
PATIENTS AND METHODS

PATIENTS

The clinical data are summarized in Table 1. All 13 patients were children from 0 to 6 years of age (median, 19 months). Eight were boys and 5 were girls. All lesions, variably described as patches, papules, nodules, or exophytic masses, were congenital and ranged in size from 0.5 to 8 cm (Figure 1A). Some lesions had increased in size with the growth of the child. All were midline lesions and all but 2 occurred on the scalp, preferentially involving the occiput and vertex. The remaining 2 lesions occurred overlying the cervical spine (C2-C3) and lumbosacral spine (Figure 1B). The “hair collar” sign, a ring of coarse hair surrounding the malformation and thought to be characteristic of cranial dysraphism, was found in 3 infants. Eleven patients had solitary defects. In each of the remaining 2 cases, 2 lesions were apparent. Other clinical abnormalities included port wine stains at a separate location in 2 patients and a congenital melanocytic nevus in patient 1. The clinical diagnoses included aplasia cutis, epidermal inclusion cyst, congenital melanocytic nevus, hemangioma, skin tag, dermal sinus tract, meningocoele, and encephalocele. Small bony defects were identifiable in 2 patients and a connecting fibrous tract to the dura was identifiable in one based on radiologic imaging results or direct surgical visualization.

AGES, VESSELS, AND MELANOCYTES WERE QUANTIFIED, AND REMNANTS OF BRAIN TISSUE WERE SOUGHT. PSAMMOMA AND COLLAGEN BODIES AND CALCIFICATION WERE ALSO SOUGHT.

THE OVERALL ARCHITECTURE VARIED IN THAT 8 LESIONS SHOWED REPLACEMENT OF BOTH THE DERMIS AND SUBCUTIS BY THE MALFORMATION, WHILE IN 4 LESIONS ONLY 1 OF THE COMPARTMENTS WAS INVOLVED. IN CASE 5 THE BIOSPY WAS TOO SUPERFICIAL TO EXCLUDE DEEP INVOLVEMENT. IN SOME SPECIMENS A LOOSE NETWORK OF PSEUDOVASCULAR SPACES IN THE SUPERFICIAL DERMIS (FIGURE 2A) CONTRASTED WITH DENSE COLLAGENOUS TISSUE CONTAINING CLUSTERS OF MELANOCYTES IN THE DEEPER PARTS. CYSTIC STRUCTURES LINED BY MENINGOTHELIAL CELLS WERE PRESENT IN 5 CASES (FIGURE 2B). THE MELANOCYTES, RECOGNIZABLE BY THEIR OVOID TO SPINDLE SHAPE WITH SCANT EOSINOPHILIC CYTOPLASM AND SMALL NUCLEI WITH FINELY STIPPLED CHROMATIN AND INDISTINCT CELL BORDERS, FORMED CORDS, STRANDS, AND SMALL NESTS AND HAD THE TENDENCY TO ENCOMPASS COLLAGEN FIBERS AND ADnexAL STRUCTURES. WE OBSERVED DECREASES AS WELL AS INCREASES IN ECCRINE GLANDS AND HAIR FOLLICLES. THERE WERE ALSO DISTORTED FOLLICLES AND AN INCREASE IN APOCRINE GLANDS AND SMOOTH MUSCLE BUNDLES. IN ONE CASE, A FOLLICULAR CYST WAS EVIDENT. PROMINENT VESSELS WERE NOTED IN 8 CASES. NECROTIC GLIAL TISSUE WAS ABSENT. CALCIFICATION WAS NOTED EITHER IN THE FORM OF PSAMMOMA BODIES (4 PATIENTS) OR IN AN UNUSUAL RETICULATE PATTERN (5 PATIENTS) (FIGURE 2C).

IMMUNOHISTOCHEMISTRY

EMA immunostaining highlighted the meningothelial cells (Figure 3) but revealed a heterogeneous pattern. Two cases failed to express EMA in the superficial aspect of the lesion but showed strong expression in the deep portion. Similarly, in specimen 5, which was transected at the level of the midreticular dermis, the loose network of pseudovascular spaces did not express EMA, but vimentin nicely outlined these spaces. CD31 and CD34, known to be vascular markers, did not react with these anastomosing channels, confirming their nonvascular nature.

Antibodies directed against S-100 protein, CD57, and CAM 5.2 failed to label meningothelial cells. S-100 was used to quantify melanocytes, which were increased at the dermo-epidermal junction in 1 specimen and within the rudimentary meningocele in 2 patients. Adjacent nerves were also identified via S-100 immunostaining in 4 patients. Anti-GFAP detected small nests of apparent glial tissue in patient 7 and also stained a few cells in patient 12. However, results of analysis for neurons with neuron-specific enolase and for axons with neurofilament in these 2 cases were negative, unlike what has been reported in most cases of heterotopic glial tissue. Meningothelial elements like those observed in both cases are not a feature of heterotopic glial tissue, but rather favor a diagnosis of rudimentary meningocele.

The pathogenesis of human neural tube defects is complex and poorly understood. Minor deviations not serious enough to significantly arrest or distort the formation of the central nervous system undoubtedly occur. Rudimentary meningocele may be this type of minor deviation and poses a problem to dermatologists in terms of diagnosis and management.

COMMENT

The 2 patients with lesions along the spine showed more complex underlying abnormalities. Both had bifid laminae and tethered spinal cords. In one of these patients, there was associated diastematomyelia and syringomyelia. The patient also had attachment of the penis to the scrotum (penis palma) with vesicoureteral reflux. In the second patient, a fibrovascular bundle pierced the dura and extended into the subdural space (Figure 1C).

All defects were locally excised. No recurrence was observed, and there was no evidence of postsurgical meningitis or cerebrospinal fluid leakage after a median follow-up of 28 months.

METHODS

Formalin-fixed and paraffin-embedded tissue from 13 skin lesions was examined in conventional and immunoperoxidase sections via a light microscope. Immunoperoxidase stains were prepared with an avidin-biotin method using monoclonal antibodies directed against epithelial membrane antigen (EMA) (Dako, Capterer, Calif), CD31 (Biogenex, San Ramon, Calif), CD34 (Novocastra, Newcastle-upon-Tyne, England), CD57 (Becton-Dickinson, San Jose, Calif), and CAM 5.2 (Becton-Dickinson), as well as polyclonal antibodies directed against S-100 protein (Dako). Two cases were evaluated further with anti-glial fibrillary acidic protein (GFAP) (Boehringer-Mannheim; Indianapolis, Ind) and vimentin (Monosan; Uden, the Netherlands).
The 13 cases of rudimentary meningocele presented herein were congenital, situated either over the scalp or midline spine. The clinical appearance was highly variable, leading to difficulty in clinical diagnosis; clinical diagnoses such as melanocytic nevus, epidermal cyst, dermal sinus tract, hemangioma, meningocele, encephalocele, and aplasia cutis were offered. Since the correct clinical diagnosis of cranial/spinal dysraphism was suspected in

<table>
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<tr>
<th>Patient</th>
<th>Age at Diagnosis/Sex</th>
<th>Size, cm/ Site of Meningocele</th>
<th>Clinical Diagnosis/Findings</th>
<th>Other Anomalies</th>
<th>Imaging Studies</th>
<th>Operative Findings</th>
<th>Follow-up Period</th>
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<tbody>
<tr>
<td>1</td>
<td>18 mo/F</td>
<td>0.8 × 0.5/ Midline occipital</td>
<td>Nevus/Nodule since birth, swelling for 6 mo</td>
<td>Congenital melanocytic nevus—right thigh</td>
<td>MRI: intact skull</td>
<td>No bone defect</td>
<td>No recurrence at 48 mo</td>
</tr>
<tr>
<td>2</td>
<td>8 mo/M</td>
<td>1.5 × 1.5/ Posterior scalp</td>
<td>Congenital nevus vs aplasia cutis/Path since birth, hair collar sign</td>
<td>None</td>
<td>Not done</td>
<td>Feeding vessel to periostium; no bone defect</td>
<td>No recurrence at 7 mo</td>
</tr>
<tr>
<td>3</td>
<td>72 mo/M</td>
<td>0.6/Posterior scalp</td>
<td>Cyst vs neural tumor/Subcutaneous nodule enlarging since birth</td>
<td>None</td>
<td>Not done</td>
<td>No bone defect</td>
<td>No recurrence at 48 mo</td>
</tr>
<tr>
<td>4</td>
<td>42 mo/M</td>
<td>3/Posterior scalp</td>
<td>Epidermal inclusion cyst/Firm tumor since birth, rapid growth by 6 y</td>
<td>Not available</td>
<td>Not available</td>
<td>Not available</td>
<td>Not available</td>
</tr>
<tr>
<td>5</td>
<td>3 mo/F</td>
<td>0.8 × 0.8/ Vertex capitis</td>
<td>Aplasia cutis/Hairless patch present since birth</td>
<td>None</td>
<td>Skull radiograph: normal</td>
<td>No bone defect</td>
<td>No recurrence</td>
</tr>
<tr>
<td>6</td>
<td>5 mo/M</td>
<td>0.9 × 0.8/Scalp</td>
<td>Congenital amelanotic nevus/Irregularly pigmented papule, growth since birth</td>
<td>Port wine stain—left arm</td>
<td>Not done</td>
<td>No bone defect</td>
<td>No recurrence at 9 mo</td>
</tr>
<tr>
<td>7</td>
<td>14 mo/M</td>
<td>1.5 × 1.5/ Cervical C2-C3</td>
<td>Skin tag, dermal sinus tract/Exophytic mass present since birth</td>
<td>None</td>
<td>MRI: cervical dermal sinus tract entering dura, tethering spinal cord</td>
<td>Bifid laminae; fibrovascular bundle piercing dura and extending to subdura</td>
<td>No recurrence at 48 mo</td>
</tr>
<tr>
<td>8</td>
<td>78 mo/F</td>
<td>1.2/Scalp</td>
<td>Congenital scalp nodule/Nodule since birth, slowly increasing in size</td>
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<td>Not done</td>
<td>No bone defect</td>
<td>No recurrence at 36 mo</td>
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<td>9</td>
<td>15 mo/F</td>
<td>1.4 × 0.9, 1 × 0.5/ Midline scalp (2)</td>
<td>Scalp masses, hemangioma/Midline scalp masses since birth</td>
<td>Port wine stain—nose</td>
<td>CT scan: no bone defect</td>
<td>No bone defect</td>
<td>No recurrence at 17 mo</td>
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<td>10</td>
<td>4 mo/M</td>
<td>4 × 2/Occipital scalp</td>
<td>Occipital encephaloceles/Tumor since birth, hair collar sign</td>
<td>None</td>
<td>MRI: postoperative changes of subcutis and calvarium; brain normal</td>
<td>0.5-cm bone defect</td>
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<td>11</td>
<td>3 wk/M</td>
<td>8 × 2.5/ Lumbosacral</td>
<td>Meningocele, diastematomyelocele/Epithelialized mass on midlumbar region, congenital</td>
<td>Spina bifida; penis palma with 1° vesicoureteral reflux</td>
<td>MRI: tethered cord, diastematomyelia, and syringomyelia</td>
<td>Bifid laminae with central bony spike; spinal cord split around bony spike</td>
<td>No recurrence at 6 mo</td>
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<td>12</td>
<td>Newborn/M</td>
<td>1.2 × 1.1/ Occipital scalp</td>
<td>Dermal sinus tract vs encephalocele/Tender nodule present since birth</td>
<td>None</td>
<td>CT scan: small soft tissue lesion with small hole in skull, dolichocephalic</td>
<td>Small bone defect with soft tissue tract to dura</td>
<td>No recurrence at 72 mo</td>
</tr>
<tr>
<td>13</td>
<td>Newborn/F</td>
<td>2 × 0.8, 0.5 × 0.5/ Vertex capitis</td>
<td>Aplasia cutis/2 Congenital bullous lesions, hair collar sign</td>
<td>None</td>
<td>CT scan: no underlying skull defect</td>
<td>No bone defect</td>
<td>No recurrence at 5 mo</td>
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*MRI indicates magnetic resonance image; CT, computed tomographic scan.
†For all of the patients the treatment was local excision.
of 1 of 13 patients, there appears to be relative unfamiliarity with these lesions. One of the most useful clinical features of cranial dysraphism, the so-called “hair collar” sign, was observed in 3 cases. Described by Commens et al, this finding consists of a collarette of hair, which encircles the malformation. This finding suggests an underlying meningocele, but the pattern is not specific, since the same clinical pattern occurs in association with an underlying encephalocele, heterotopia of brain tissue, or membranous aplasia cutis. Membranous aplasia cutis refers to congenital, sharply margined ovoid scalp defects. In contrast to rudimentary meningocele, this condition often presents as multiple lesions with a tendency to regress from an initial cystic or bullous appearance to a scar. Despite these differences, there seems to be an overlap with rudimentary meningocele in terms of the clinical picture. One of our patients presented with 2 bullous scalp defects that could not be differentiated from membranous aplasia cutis on clinical grounds. The histopathologic features in our patient, however, were clearly those of rudimentary meningocele based on the presence of meningeal tissue, which by definition is not found in membranous aplasia cutis.

In view of the relative rarity of rudimentary meningocele, the histopathologic diagnosis is difficult. The microscopic features are often subtle, with meningeal tissue simulating the appearance of vascular or connective tissue. Meningocytes tend to encircle collagen bundles and are sometimes accompanied by psammoma bodies; these signs should be sought as clues to the microscopic diagnosis. An additional helpful clue was a decrease or increase in adnexal structures, similar to the pattern that can be seen in some forms of epidermal nevi and aplasia cutis. Cystic structures lined by meningotheelial cells, as would be seen in a classic meningocele, were occasionally evident, highlighting the similarity between the classic and rudimentary forms of this disorder. Classic meningocele can be separated from rudimentary meningocele only on the basis of clinical data. In general, meningoocytes were few in number, were positioned between collagen bundles, and created a microscopic pattern resembling vascular spaces. These pseudovascular spaces can easily be misinterpreted as a vascular neoplasm such as lymphangioma or even angiosarcoma. Apart from the clinical picture, absence of nuclear pleomorphism and mitotic figures, and the lack of reactivity to vascular markers, such as CD31 and CD34, should enable exclusion of a vascular neoplasm.

Another histopathologic finding that could contribute to error in microscopic diagnosis is the presence of syncytial giant cells. These cells result from fusion of meningoocytes and may simulate the pattern of giant cell fibroblastoma, a form of dermatofibrosarcoma protubersans usually observed in children. Myxoid stroma noted in either case may further obscure the diagnosis. Immunoperoxidase staining is vital in distinguishing these disorders, as dermatofibrosarcoma protuberans typically expresses CD34 and fails to express EMA, while rudimentary meningoceles display opposite reactivity. Although EMA expression is vital to the identification of cells of meningotheelial lineage, it is important to note that EMA failed to label the network of pseudovascular spaces located in the superficial dermis in 3 of our patients. Only thorough examination revealed EMA-positive meningoocytes in the deeper parts of 2 specimens. Therefore, in specimens from superficial biopsies that do not extend below the level of the middermis, as was the case in patient 5, meningotheelial elements may not be detectable. With respect to superficial biopsies, some cases de-
scribed as membranous aplasia cutis with no meningeal or brain tissue detected by immunohistochemistry may actually be examples of rudimentary meningocele.15 We observed only focal EMA positivity in normal leptomeningeal tissue, as reported by others.8,20,21 The diffuse meshwork of subarachnoid cells shares many features with the loose network seen in rudimentary meningocele, and this meshwork does not label at all with EMA. An alternative explanation is that the dilution of the EMA antibody that usually works well with epithelial cells might not work as well in cells of meningothelial lineage.

Speculation about the etiology of rudimentary meningocele started with the first observation of this malformation by Winkler in 1904, in which he described a "peculiar disease of the skin and subcutaneous fat." An increasing number of reports of this entity have appeared in the more recent literature, and with them a variety of divergent opinions on the pathogenesis have been suggested. Some authors consider these lesions to be a form of meningocele with an obliterated intracranial communication, while others refer to them as remnants of the neural crest. A more historical theory suggests that they might represent intradermal nevi containing psammoma bodies.3,7,16,22-26 In addition, these lesions were also postulated to arise from the sheath cells of cutaneous nerves or to reflect continued growth of meningeal cells within a perineural environment.8,27

A fairly new theory about closure of the neural tube may yield greater insight into the pathologic mechanism of rudimentary meningocele, as it seems to explain some of the clinical findings.10,11 Traditionally, neural tube closure was believed to begin in the cervical region and to proceed from there in a continuous bidirectional way.10 (Figure 4A). More recent observations in humans and animals, however, indicate multisite initiation of neural tube closure10 (Figure 4B). The failure of closure to take place partially or completely would thus result in neural tube defects.11 Mouse models have shown 4 separate sites of neural tube fusion. Homologies of early embryogenesis in mice and humans as well as previous

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<th>Case</th>
<th>Meningothelial Elements in Superficial/Deep Reticular Dermis</th>
<th>Psammoma Bodies/ Collagen Bodies</th>
<th>Calcification Pattern</th>
<th>Skin Appendages</th>
<th>Vessels</th>
<th>Melanocytes</th>
<th>Nerves</th>
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*− indicates absent; +, present; ++, abundant; PS, psammoma body; EG, eccrine gland; HF, hair follicle; ↓, decreased DEJ; dermoepidermal junction; NL, normal; RC, reticulate calcification; AG, apocrine gland; ↑, increased; ellipses, not applicable; and RM, rudimentary meningocele.
genes control individual closure sites. Interestingly, fa-
defects is genetic susceptibility, with a risk of repetitive
among two siblings. Simple displacement of menin-
gocytes along peripheral nerves is an unlikely alternate
illustrations and photographs of human embryos sug-
gest that neural tube fusion occurs at sites similar to those in
mice. Of particular interest are the locations of such neural
tube defects, as they coincide with the distribution
observed in our cases of rudimentary meningocele (Figure 4C). Closure in the occipital area appears to be
achieved by a membrane rather than a midline fusion of
folds, which explains why such defects may or may not
appear in the midline. Another feature of neural tube
defects is genetic susceptibility, with a risk of repetitive
events in affected families. This suggests that specific
genes control individual closure sites. Interestingly, fa-
miliar occurrence of rudimentary meningocele has been
noted in an autosomal dominant inherited pattern and
among two siblings. Simple displacement of menin-
gocytes along peripheral nerves is an unlikely alternate
explanation for such observations.

Another finding that supports classifying rudimen-
tary meningocele as a form of meningocele is the iden-
tification of connections, albeit rudimentary ones, that
extend from the lesion to dura, and in some instances
are associated with minute bony defects. We were
able to detect such fibrous tracts in 2 cases and a small
osseous defect in 1 infant.

In light of the likely pathogenesis of these lesions, prior
to complete surgical excision, we believe that imaging
studies should be obtained to exclude the possibility of a
communication to the central nervous system. If there is evidence of an underlying connection, the patient should be
referred for neurosurgical evaluation. If a communication to the central nervous system can be excluded, simple ex-
cision would be appropriate, and neither recurrence nor neurologic consequences would be expected to occur.

The pathologic mechansism of rudimentary menin-
goceles remains enigmatic and has not been completely elucidated. However, our clinical and histopathologic find-
ings and those of other investigators support the conclu-
sion that this entity is a form of dysraphism. We be-
lieve this process is a non-neoplastic condition, and thus
the designation of cutaneous meningioma should not be
used as a synonym for rudimentary meningocele.

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