Congenital inclusion cyst of the subgaleal space

A. Basit Chaudhari, M.B., B.S., F.R.C.S., F.M.C.S. (Nig.),
Foluso Ladao, M.D., Ch.B., B.O.A.O., D.M.R.D.,
Victor P. N. Mordi, M.B., B.S., M.D.,
Kalyanpur J. Choudhury, M.D., M.N.A.M.S.,
Attia Naseem, M.B., B.S., M.D., and Joan A. Obe, M.D.

Departments of Surgery, Radiology, Morbid Anatomy, and Anesthesia, College of Medicine and Lagos University Teaching Hospital, Lagos, Nigeria, and Departments of Pathology, Mount Vernon Hospital, Mount Vernon, and Montefiore Hospital, Bronx, New York

An unusual type of congenital subgaleal cyst, either dermoid or epidermoid, was found in 21 Nigerian infants. This entity was located in the midline of the scalp, anywhere from above the nasion to the inion, and occupied the subgaleal space. These cysts were noted soon after birth and gradually enlarged. They had no intracranial extension and were easily excised intact. Clinically, radiologically, and histologically they were similar to congenital inclusion dermoid cysts of the anterior fontanel. The histology and some of the peculiarities of these cysts are described. Although most of these cysts occur at the anterior fontanel, they can occur anywhere in the midline, and the subgaleal space of the anterior fontanel is not the exclusive site, as has been claimed by previous authors.

Key Words: dermoid cyst, epidermoid cyst, midline scalp cyst, anterior fontanel cyst, congenital inclusion cyst, subgaleal space

Congenital dermoid or epidermoid cysts occur at the midline or at the lines of embryonal fusion. Ectopia or heterotopia of skin tissue may occur between 3 and 5 weeks of intrauterine life. This is the usual time of development of ectopic skin tissue in deeply situated dermoid cysts along the craniospinal axis; however, laterally situated intracranial epidermoid cysts are postulated to occur at a later stage of embryogenesis. At even later stages, inclusion or ectopic tissue may occupy more superficial structures such as bone, pericranium, or subgaleal space. The strata occupied by dermoid or epidermoid cysts merely reflect the stage of embryogenesis at which the biological process of inclusion of ectopic cutaneous tissue took place. Subgaleal cysts result from inclusion of sequestrated epithelial remnants or ectodermal displacement in the subgaleal space.

We are reporting the unusual features of congenital inclusion dermoid cysts of the subgaleal space in 21 Nigerian children over a period of 6 years from 1975 to 1981. The fact that these cysts can occur in the subgaleal space anywhere in the midline of the scalp is particularly stressed. As far as we know, this is the only series in the English literature that describes a cyst location other than at the anterior fontanel.

Summary of Cases

The clinical, radiological, and operative findings in these infants are summarized in Table 1. The age at the time of clinical diagnosis ranged from 2 1/2 weeks to 2 1/2 years and females predominated in a ratio of 2:1 (14 females and seven males). The most important factor precipitating admission to the hospital was apprehension in regard to the nature of the lesion in the minds of the parents or attending clinician. Pediatricians in our teaching institutions in Nigeria are well aware of this entity, and in 95% of the cases a correct diagnosis had been made when the cases were referred through them.

Clinically, these cysts resembled encephaloceles, but there was no evidence, either radiological or at operation, of any demonstrable intracranial connection or extension. The cysts were solitary and variable in size; they were generally found in the subgaleal space of the anterior fontanel, but two cysts were situated in front of it and one each at its anterior and
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FIG. 1. The subgaleal cysts in this series were located: well behind the anterior fontanel (frontal view, A; lateral view, B), over the external occipital protuberance (C), and at the anterior fontanel (the classical location, D).

posterior angles. One lay over the vertex well behind the anterior fontanel (Fig. 1A and B), and another was located over the occipital protuberance (Fig. 1C). The remaining 15 cysts were situated in the classical position at the anterior fontanel (Fig. 1D).

The cysts were slightly mobile, and all those that contained relatively clear fluid transilluminated well. The lesions had either been present since birth or were noted soon after, and were slowly enlarging. They were soft, fluctuant, and nontender, with a sessile base. There was a feeble transmission of cough impulse with increased tension in some of the patients in whom the anterior fontanel was still open. The cysts varied in diameter from 1.5 to 5 cm, and were well covered with intact skin.

Skull films revealed only a soft tissue mass overlying intact bone which was slightly indented. In some of the cases, air cystography was performed after aspiration of the fluid. This study delineated the sac and defined its limits, and clearly showed that there was no escape of air into the intracranial cavity, thus confirming the entirely extracranial location of the cyst (Fig. 2). Air ventriculography and cisternal pneumoencephalography were not done routinely.

The diagnosis was made largely on clinical and plain radiographic findings. Air cystography carries the risk of infection and it was limited to cases in which further elaboration was required. We do not recommend it as a routine procedure. The treatment of choice was operative excision of the cyst through a transverse or elliptical skin incision across the summit of the cyst.

At operation, a well encapsulated, translucent cyst with a variable amount of collagen in the capsule could easily be dissected from the loose areolar tissue and shelled out intact with no recurrence. In none of the cases was there any connection with the intracranial cavity or overlying skin.

The smaller cysts generally, but not always, contained clear colorless fluid, with occasional caseous, cheesy-white floating particles. Some of the smaller cysts and most of the larger cysts contained fluid that varied in color and consistency from straw-colored, buttery yellow, yellowish-brown, or grayish with occasional hairs. The clear fluid of most of the smaller cysts was low in protein, cholesterol, and sugar content, of low pH with low sodium and chloride con-
## TABLE 1

**Congenital inclusion cyst of the subgaleal space**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, Sex</th>
<th>Description of Cyst</th>
<th>Diameter (cm)</th>
<th>Location</th>
<th>Cyst Contents On Aspiration</th>
<th>Cyst Contents At Removal</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2½ wks, F</td>
<td>soft, cystic, present since birth</td>
<td>2.5</td>
<td>anterior fontanel</td>
<td>clear fluid</td>
<td>clear fluid, some caseous nonsmelling material, buttery-yellow fluid</td>
</tr>
<tr>
<td>2</td>
<td>3 wks, F</td>
<td>soft, cystic, fluctuant</td>
<td>3</td>
<td>anterior fontanel</td>
<td>clear fluid</td>
<td>pearly-gray crystals on cyst wall</td>
</tr>
<tr>
<td>3</td>
<td>4 wks, F</td>
<td>soft, cystic, fluctuant</td>
<td>4</td>
<td>anterior fontanel</td>
<td>clear fluid</td>
<td>crumbly yellow crystals on cyst wall</td>
</tr>
<tr>
<td>4</td>
<td>5 wks, M</td>
<td>soft, cystic, fluctuant</td>
<td>2.5</td>
<td>anterior fontanel</td>
<td>yellowish, clear fluid</td>
<td>fine granular material on cyst wall</td>
</tr>
<tr>
<td>5</td>
<td>7 wks, F</td>
<td>soft, cystic, enlarging since birth</td>
<td>3</td>
<td>anterior fontanel</td>
<td>clear fluid</td>
<td>clear fluid, caseous whitish material</td>
</tr>
<tr>
<td>6</td>
<td>4 mos, M</td>
<td>soft, cystic; recurred after aspiration</td>
<td>3</td>
<td>anterior fontanel</td>
<td>clear fluid</td>
<td>turbid, yellowish-white fluid, brown crystals on cyst wall</td>
</tr>
<tr>
<td>7</td>
<td>5 mos, M</td>
<td>soft, cystic, fluctuant</td>
<td>3.5</td>
<td>anterior fontanel</td>
<td>butty yellow</td>
<td>clear fluid with caseous particles floating</td>
</tr>
<tr>
<td>8</td>
<td>2½ yrs, F</td>
<td>soft, cystic, fluctuant, enlarging since birth</td>
<td>3.5</td>
<td>anterior to anterior fontanel</td>
<td>clear fluid</td>
<td>turbid fluid with grayish crystals on cyst wall</td>
</tr>
<tr>
<td>9</td>
<td>6 mos, M</td>
<td>soft, cystic, enlarging since 10 days old</td>
<td>3</td>
<td>anterior angle of anterior fontanel</td>
<td>yellowish-brown fluid</td>
<td>clear fluid with caseous material floating</td>
</tr>
<tr>
<td>10</td>
<td>7 mos, M</td>
<td>soft, cystic, fluctuant, enlarging since birth</td>
<td>3</td>
<td>anterior fontanel</td>
<td>clear fluid (air study)</td>
<td>yellow fluid</td>
</tr>
<tr>
<td>11</td>
<td>10 mos, F</td>
<td>soft, cystic, fluctuant</td>
<td>3</td>
<td>anterior to anterior fontanel</td>
<td>yellowish-brown fluid</td>
<td>turbid fluid with soft granular material</td>
</tr>
<tr>
<td>12</td>
<td>11 mos, M</td>
<td>soft, cystic, fluctuant</td>
<td>2.5</td>
<td>occipital protuberance</td>
<td>—</td>
<td>clear fluid with caseous particles floating; pearly white material on cyst wall</td>
</tr>
<tr>
<td>13</td>
<td>2½ yrs, F</td>
<td>soft, cystic</td>
<td>4</td>
<td>posterior angle of anterior fontanel</td>
<td>clear fluid</td>
<td>clear fluid, some caseous material</td>
</tr>
<tr>
<td>14</td>
<td>1 yr, F</td>
<td>soft, cystic</td>
<td>2.5</td>
<td>anterior fontanel</td>
<td>infected, clostridium B*</td>
<td>yellowish turbid fluid</td>
</tr>
<tr>
<td>15</td>
<td>1½ yrs, F</td>
<td>soft, cystic, fluctuant</td>
<td>3.5</td>
<td>anterior fontanel</td>
<td>—</td>
<td>yellow-brown fluid with grayish crystals floating &amp; resting against cyst wall</td>
</tr>
<tr>
<td>16</td>
<td>2 yrs, F</td>
<td>soft, cystic, fluctuant</td>
<td>5</td>
<td>anterior fontanel</td>
<td>—</td>
<td>clear fluid with caseous material floating</td>
</tr>
<tr>
<td>17</td>
<td>11 mos, F</td>
<td>soft, cystic, fluctuant</td>
<td>4</td>
<td>anterior fontanel</td>
<td>—</td>
<td>buttery-yellow fluid, amorphous crumbly crystals on cyst wall</td>
</tr>
<tr>
<td>18</td>
<td>1½ yrs, F</td>
<td>soft, cystic, fluctuant</td>
<td>3</td>
<td>anterior fontanel</td>
<td>—</td>
<td>yellowish-brown thick fluid</td>
</tr>
<tr>
<td>19</td>
<td>1 yr, M</td>
<td>soft, cystic, fluctuant</td>
<td>3.5</td>
<td>anterior fontanel</td>
<td>—</td>
<td>clear fluid with caseous material floating</td>
</tr>
<tr>
<td>20</td>
<td>6 mos, F</td>
<td>soft, cystic, fluctuant, relatively mobile</td>
<td>3</td>
<td>well behind anterior fontanel, at vertex anterior fontanel (rather deeply embedded)</td>
<td>clear fluid (air study)</td>
<td>clear fluid (air study)</td>
</tr>
<tr>
<td>21</td>
<td>5 mos, F</td>
<td>soft, cystic, fluctuant</td>
<td>2.5</td>
<td>anterior fontanel</td>
<td>clear fluid</td>
<td>clear fluid with caseous material floating; pearly gray crystals on cyst wall</td>
</tr>
</tbody>
</table>

* Aspiration had previously been attempted at another institution.

Contents, and relatively high potassium. The colored fluid was alkaline (with a high pH) with high sodium, cholesterol and chloride content, and relatively low potassium. The fluid was bacteriologically sterile except in Case 14, in which *Escherichia coli* was isolated (Table 1).

On macroscopic and histological examination, the cysts were reported as epidermoid or dermoid cysts. The sections showed cystic cavity lined by keratinizing squamous epithelium containing keratin debris in the epidermoid lesions. Skin appendages, including hair follicles, and sebaceous and sweat glands, were seen in the fibrous element of the cyst wall in the dermoid cysts.

### Discussion

Odeku\(^\text{12-14}\) and Adeloye\(^\text{1}\) deserve credit for describing subgaleal cysts of the anterior fontanel with clarity and precision. The previous accounts were sketchy and vague and often one could draw information only from indirect inference from the pictorial descriptions. Even then the nature of the lesions was not consistent.\(^\text{1}\) Jackson and Thompson\(^\text{6}\) gave a pictorial description of a small sessile frontal encephalocele (often confused with a sebaceous cyst of the scalp) in a child who looked normal as far as could be discerned. Matson\(^\text{10}\) illustrated a congenital inclusion epidermoid at the anterior fontanel which extended through both layers...
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of the cranium but did not penetrate the dura in a boy of 20 months (his Fig. 468). No further mention of it was made in the text.

Intracranial and intraspinal dermoid cysts have been known since the time of Cruveilhier in 1829, and several accounts and series have been reported since then.7-9,11,17,18 Extracranial inclusion dermoid cysts of the subgaleal space have been recognized recently, but they are still regarded as rare.15 Most of the series have emanated from various African centers.1,5,12-14,16 The salient features and peculiarities of this type of cyst are its location in the subgaleal space of the anterior fontanel, the female preponderance, the prevalence in the negroid race, and specific cyst fluid composition.1,5,12-14,16 Odeku termed it a "peculiar subgaleal inclusion cyst."

All previous accounts suggest that subgaleal inclusion cysts are always situated in the subgaleal space of the anterior fontanel, and perhaps this was thought to be the most peculiar feature of this cyst.1,5,12-14 The anterior fontanel is one of the most important sites of embryonal fusion in the cranium and thus constitutes a kind of strategic point to attract the ectopic cutaneous tissue. Although our series also showed a prevalence at the anterior fontanel, this cyst can occur anywhere in the midline, thus supporting the reported occurrence of dermoid and epidermoid cysts at the midline or line of embryonal fusion.1

It is difficult to explain the female preponderance and the prevalence in the negroid race. Perhaps this incidence is related to a structural difference in the scalp. This cyst is known to occur in other races but, according to previous series, the majority were in the negroid race. Our series supported this finding. The female to male ratio in our series corresponded with that of previous series (2:1).

There appeared to be some relationship between the size of the cyst and character of its fluid contents. Cyst fluid formation appeared to be the active secretion of sweat and sebaceous glands found in the lining of squamous stratified epithelium. The biochemical analysis of the fluid varied according to the number of sebaceous and sweat glands and the mechanism of their secretion, whether the gland was apocrine, eccrine, or holocrine. Straw- or yellow-colored fluid which was otherwise clear probably was the result of chromidrosis, that is, the secretion of pigmented sweat by the apocrine sweat glands. This phenomenon is rarely observed in the scalp when vestigial apocrine glands are functional.2 The electrolyte contents were not always similar to that of sweat, which may be due to dilution by sebaceous secretion or some kind of mechanism of differential adsorptions of electrolytes on cholesterol crystals and the soft lamellae formed by the progressive desquamation and breakdown of keratin from the epithelial lining of the cyst.

Malignant change has been recorded in dermoid and epidermoid cysts of the craniocephalic axis in cases of incomplete removal.4,5,17 None was seen in our series or previous reports.1,5,12-16 probably due to total excision at operation. Cases recorded in adults in which the operation was postponed showed no evidence of malignancy.15

The defect in the external table of the calvaria underneath the subgaleal cyst was variable in depth. There was no direct relationship between the depth of the bone defect and the duration of the cyst. This appears to be a part of the embryological anomaly. There were examples in infants under 6 months of age who had very pronounced depression in the bone as compared to patients over 2 years old. Also, when operation was postponed for some months, a comparison of the x-ray films at different times before excision showed no increase in the size of the cavity, indicating that the bone defect did not result from downward pressure. This fact favors the hypothesis that the strata that are occupied by subgaleal cysts, in common with dermoid or epidermoid cysts elsewhere, merely reflect the stage of embryogenesis at which the biological process of inclusion of ectopic cutaneous tissue took place.

When a subgaleal cyst is at its classical location (at the anterior fontanel), it is quite simple to diagnose. However, cysts at other locations suggest a meningocele, encephalcele, sebaceous cyst, lymphangioma, hemangioma, cystic hygroma, or cephalhematoma, but inclusion cysts of the subgaleal space should be considered in the differential diagnosis.

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References


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Address reprint requests to: Professor A. Basit Chaudhari, % Dr. K. Mustafa, Bedford Farms, McLain Street, Mount Kisco, New York 10549.