A morphological classification of sincipital encephalomeningoceles

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Craniocerebral defects and cerebral abnormalities as revealed by postmortem dissection in 12 patients with sincipital encephalomeningocele are reported. The various methods of classifying this lesion are discussed. A classification based on the location of the defect in the cranium is outlined. The clinical application of such a classification and its usefulness in the surgical management are emphasized.

**Key Words**  sincipital encephalomeningocele

Meningoceles in the anterior part of the head are rare in Western Europe and America but appear to be common in Southeast Asia. We have found 100 cases in the past 8 years and have reported the clinical manifestations and treatment elsewhere. We have done postmortem dissection on 12 patients with the herniation in the region from the forehead to the nose; our findings, we believe, throw some light on the unsettled classification of such lesions. This communication is an attempt to make a pragmatic and useful classification based on the location of the defect in the cranium.

The results of postmortem dissection in 12 patients, indicating the site of the facial mass and the skull defect, are shown in Fig. 1, and the findings in the patients are tabulated in Table 1. Based on the location of the defect in the skull, we have divided them into three groups, as follows:

1. Fronto-ethmoidal encephalomeningocele
   A. Nasofrontal (Cases 1–6)
   B. Naso-ethmoidal (Cases 7–9)
   C. Naso-orbital (Case 10)

2. Interfrontal encephalomeningocele (Case 11)
3. Craniofacial cleft (Case 12).

**Fronto-ethmoidal Encephalomeningoceles**

In 10 patients, the opening of the defect in the skull as seen intracranially was located at the junction between the frontal and ethmoidal bones. Nine had a single opening in the midline corresponding to the site of the foramen cecum. The crista galli was situated at the posteroinferior rim of the defect. One patient (Case 10) had two openings at the anterior end of the cribriform plate, one on each side. The crista galli was at the posterior part of the bridge of the bone between the two openings. The patients in this group can be divided into three subgroups according to the site of the facial end of the orifice (Fig. 2).

**Nasofrontal Type (Cases 1 through 6)**

The skull defect as seen from the facial aspect was almost identical in these six patients; it was round or ovoid at the bregmatic
region between the deformed orbits (Fig. 3). The crista galli projected into the defect from its inferior rim. The anterior portion of the medial orbital wall was displaced laterally. The ethmoidal bone forming the middle portion of the floor of the anterior cranial fossa was low in position compared with the roof of the orbits. When seen intracranially, the anterior cranial fossa was unusually deep in the middle. The nasal bones and the frontal processes of the maxillary bone, as well as the nasal cartilage, were in normal relationship. The cranial defect was therefore between the frontal and ethmoidal bones when seen from the intracranial side and between the frontal and nasal bones as seen from the outside. The two orifices were, however, very close together.

All patients in this subgroup had a mass at the glabella or at the root of the nose (Fig. 4), the size and character of which varied greatly. The size ranged from 2 cm in diameter to one larger than the child's head. Two patients had no skin covering and brain tissue presented in the form of two nodular masses separated by a median constriction, corresponding to both frontal tips and the intervening falx cerebri. The mass in the other

Fig. 1. Drawings to illustrate the findings in all 12 patients. The appearance and location of the mass are shown in the upper row, the defects of the bones of the face in the middle row. The intracranial openings in Cases 1 through 10 and the defects of the cranial vault in Cases 11 and 12 are shown in the lower row.
TABLE 1

Details of findings on dissection of 12 encephalomeningoceles

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at Death</th>
<th>Sex</th>
<th>Location of Mass</th>
<th>Size of Mass (cm)</th>
<th>Skin Covering &amp; Wall of Sac</th>
<th>Location &amp; Size (cm) of Cranial Defect*</th>
<th>Amount of Herniated Brain</th>
<th>Brain</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8 days M</td>
<td></td>
<td>glabella</td>
<td>4 X 3 X 2</td>
<td>bleeding, no skin covering, thin wall of sac</td>
<td>FE 2 X 2.5 NF</td>
<td>large</td>
<td>holotelencephaly, microgyria &amp; agyria</td>
</tr>
<tr>
<td>2</td>
<td>1 mo. M</td>
<td></td>
<td>glabella</td>
<td>2 X 2 X 5</td>
<td>thin &amp; shiny skin thin wall of sac</td>
<td>FE 2.5 X 2.5 NF</td>
<td>small (2 X 2 X 3 cm)</td>
<td>small frontal lobes, long quadrigenimal plate &amp; aqueduct</td>
</tr>
<tr>
<td>3</td>
<td>1 mo. F</td>
<td></td>
<td>glabella</td>
<td>4 X 3 X 2</td>
<td>no skin covering, infected, thin wall of sac</td>
<td>FE 2 X 2 NF</td>
<td>large</td>
<td>left unilateral hydrocephalus, elongated brain stem &amp; hypothalamus</td>
</tr>
<tr>
<td>4</td>
<td>prem, F newbn.</td>
<td></td>
<td>glabella</td>
<td>4 X 3 X 2</td>
<td>no skin covering, bleeding, no sac wall</td>
<td>FE 2 X 2 NF</td>
<td>moderate</td>
<td>small frontal lobes, traction of cortex &amp; anterior communicating artery forward</td>
</tr>
<tr>
<td>5</td>
<td>2 mos. F</td>
<td></td>
<td>glabella</td>
<td>5 X 3 X 2</td>
<td>ulcerated, infected, thin wall of sac</td>
<td>FE 3 X 3 NF</td>
<td>moderate (3 X 4 X 4 cm)</td>
<td>right unilateral hydrocephalus, reverse direction of optic nerve &amp; internal carotid artery, long brain stem, abnormal position of falx cerebri</td>
</tr>
<tr>
<td>6</td>
<td>2 mos. M</td>
<td></td>
<td>glabella</td>
<td>10 X 10 X 15</td>
<td>thin &amp; shiny skin, thin wall of sac</td>
<td>FE 4 X 2 NF</td>
<td>about one-half</td>
<td>fairly well preserved cortical gyri, stretched cortex &amp; brain stem</td>
</tr>
<tr>
<td>7</td>
<td>3 mos. F</td>
<td></td>
<td>wide base of nose with right mass</td>
<td>0.5 X 1 X 1.5</td>
<td>thin skin over mass, shiny at cyst</td>
<td>FE 2 X 2 NE</td>
<td>small (1 X 2 X 2 cm)</td>
<td>bilateral hydrocephalus, elongated brain stem</td>
</tr>
<tr>
<td>8</td>
<td>2 yrs F</td>
<td></td>
<td>both sides of base of nose</td>
<td>2 X 1 X 1</td>
<td>thin skin over mass, shiny at cysts</td>
<td>FE 2 X 3 NE</td>
<td>moderate (2 X 2 X 4 cm)</td>
<td>right unilateral hydrocephalus, elongated brain stem, abnormal position of falx cerebri</td>
</tr>
<tr>
<td>9</td>
<td>1½ yrs. M</td>
<td></td>
<td>both sides of wide base of nose</td>
<td>4 X 3 X 2</td>
<td>normal skin with scar, thin wall of sac</td>
<td>FE 2 X 4 NE</td>
<td>moderate (2 X 4 X 4 cm)</td>
<td>bilateral hydrocephalus, elongated brain stem</td>
</tr>
<tr>
<td>10</td>
<td>1 mo. M</td>
<td></td>
<td>both infra-orbital</td>
<td>2 X 1 X 0.5</td>
<td>normal skin, thick wall of sac</td>
<td>FE; NO 0.7 X 0.7 left 0.4 X 0.2 right</td>
<td>minimal (olfactory bulb)</td>
<td>normal</td>
</tr>
<tr>
<td>11</td>
<td>8 days F</td>
<td></td>
<td>midforehead</td>
<td>7 X 5 X 2</td>
<td>normal skin, thick wall of sac</td>
<td>interferfrontal 6 X 7</td>
<td>moderate (2 X 6 X 6 cm)</td>
<td>normal cortex, constricted ring at frontal lobes</td>
</tr>
<tr>
<td>12</td>
<td>newbn. F</td>
<td></td>
<td>whole head</td>
<td>15 X 10 X 8</td>
<td>thin &amp; shiny, right normal, left</td>
<td>craniofacial 5 X 6</td>
<td>more than half</td>
<td>severe damage, partly macerated, two constrictions at cerebrum</td>
</tr>
</tbody>
</table>

* FE = fronto-ethmoidal, NF = nasofrontal, NE = naso-ethmoidal, NO = naso-orbital

two patients had a thin skin covering which had ruptured and become infected. The last two patients had cystic masses covered with thin and shiny skin.

Upon dissection of the brain, one patient (Case 1) had a single cerebrum without a falx cerebri or corpus callosum, that is, a holotelencephaly (Fig. 5). The remaining five patients had herniation of both frontal tips and the intervening falx cerebri into the sac. The amount of the herniated brain varied from the size of a fingertip to almost half of the brain (Fig. 6). When a large amount of brain was herniated, the cortical gyri and sulci converged at the neck of the opening. Certain areas of cortex were smooth with faint or no convolutions. The cerebral blood vessels also followed the brain into the herniation. In one patient, the anterior communicating artery was displaced to the level of the crista galli (Fig. 7). The olfactory apparatus was almost invariably herniated and the ol-
tactory tract stretched. The brain stem was affected to a variable degree. In some, there was an elongation of the quadrigeminal plate and angulation of the aqueduct, which could very well be the cause of hydrocephalus. In others, the brain stem and hypothalamus were elongated to unrecognizable structures. In severe cases, the optic nerves, after passing out of the optic canal, bent sharply forward and took a forward course to the inferior surface of the brain as far as 1 cm away. The internal carotid arteries followed the same course as the optic nerves. In one patient, the tip of the temporal lobe was seen to herniate above the sphenoid ridge into the anterior cranial fossa. The frontal tips were not equally herniated, and there was some

**Fig. 3.** Case 1. Nasofrontal subgroup of fronto-ethmoidal encephalomeningocele showing the defect between the frontal bone on one side and the ethmoidal and nasal bones on the other. The crista galli is seen protruding into the defect. Note the low position of the cribriform plates in relation to the roof of the orbits. This patient is Case 5 in our previous report.3

**Fig. 4.** Case 4. Nasofrontal subgroup of the fronto-ethmoidal encephalomeningocele showing the typical location of the mass in this subgroup.
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rotation of the brain. Unilateral hydrocephalus was seen in two of these five patients. The herniated portion of the brain underwent variable degrees of change, from one that was rather well preserved to one that had become a firm nodular mass of adherent scar.

In one patient (Case 5) the falx cerebri attached to the tentorium cerebelli along the left apex of the petrous pyramid instead of the median portion. The right cerebral hemisphere, which was hydrocephalic, was larger than the left one and occupied the right half as well as the posterior portion of the left half of the cranial cavity. The superior sagittal sinus also deviated to the left and lay close to the point of attachment of the falx cerebri.

Naso-ethmoidal Type (Cases 7 through 9)

The cranial defect in three patients was between the nasal bones and the nasal cartilage as seen from the face. The nasal bones and a portion of the frontal process of the maxillary bone were attached to the frontal bone above the sac of the encephalomeningocele, forming the anterosuperior wall of the canal for the sac. The posteroinferior wall was formed by the nasal cartilage and nasal septum which attached superiorly to the ethmoidal bone. The crista galli was usually seen on top of the ethmoidal bone in the depths of the hole. The lateral wall, which at times was membranous instead of bony, was formed by the medial wall of the orbit (Fig. 8).

In this subgroup, the neck of the encephalomeningocele was long. Its intracranial orifice was between the frontal and ethmoidal bones, and its outer end lay between the nasal bones and the nasal cartilage. The distance between the nasal bones and the nasal

Fig. 5. Case 1. Superior view (left) and coronal section (right) of the brain in an 8-day-old boy with a nasofrontal encephalomeningocele showing holotelencephaly.

Fig. 6. Case 6. Brain showing constriction in the middle corresponding to the neck of a nasofrontal encephalomeningocele. About one-half of the brain was outside the cranial cavity. Note the rather well-formed cortical gyri.
group A. All three had a moderate degree of hydrocephalus. In one (Case 8), the falx cerebri was attached along the apex of the left petrous pyramid (similar to Case 5). In another, there was agenesis of the corpus callosum, which had been replaced by an interhemispheric cyst, measuring $6 \times 3 \times 1.5$ cm, containing cerebrospinal fluid.

**Naso-orbital Type (Case 10)**

This patient had two masses, one on each side, at the nasolabial fold between the nose and lower eyelids. They were rather firm with normal skin covering. Upon dissection of the face, thick-walled cystic masses were found, at the depth of which small nodules of the herniated brain were located (Fig. 11). The frontal and nasal bones and cartilage were in normal relationship. The frontal process of the maxillary bone was defective in the middle, forming the anterior limits of cartilage varied. When they were in close proximity, the sac of the encephalomeningocele was localized within the widened bridge of nose. In one patient, it extended laterally and the lesion almost resembled the naso-orbital type to be described.

The mass in these three patients was at the root of the nose extending to the inner canthus of the eyes. One of them had cystic swellings on both sides of the root of the nose (Fig. 9). Another patient had diffuse swelling of the bridge of the nose, with normal skin covering. The mass was rather firm in the middle and soft on both sides. In the third patient, a shiny cystic mass was present to the right of the midline on top of the wide bridge of the nose.

On dissection of the face, it was found that the cystic swellings and the soft areas corresponded to the herniated frontal tips and an overlying space containing cerebrospinal fluid. In the midline, there was a thick fibrous septum which was a prolongation of the falx cerebri (Fig. 10).

The brains of these patients showed changes similar to those described in Sub-
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Inferior margin, a 1 cm metopic suture was seen extending downward to join the frontonasal suture. Posteriorly the defect merged into the anterior fontanel. The sagittal and coronal sutures were in normal positions. The rest of the skull showed the so-called "beaten-silver" appearance. Both of the frontal lobes extended into and filled the frontal swelling. A constriction ring could be seen on the cerebral cortex corresponding to the rim of the bone defect. The ventricles were slightly enlarged but the brain was otherwise normal.

Craniofacial Cleft

We have seen two patients with this malformation of the head and face. One of them (Case 12) was studied by postmortem dissection. The child had a small skull with a big hole on the top. Two large cystic masses extended up from the area of the bone defect. A complete cleft lip and palate was seen on the left. The right nostril was displaced rostrally and was situated close to the base of the external orifice. The lacrimal bone and the lamina papyracea of the ethmoid constituted the posterior edge of the external orifice. Intracranially, two holes were seen at the foremost end of the cribriform plate just in front of the crista galli. They measured about 6 mm on the right and 4 mm on the left. In this patient, the canal forming the neck of the encephalomeningocele was very long. The brain was globular in shape and was within normal limits except for the herniation of the tips of both olfactory bulbs and also, on the left side, the surrounding orbital cortex.

Interfrontal Encephalomeningocele

One patient (Case 11) was born with multiple anomalies: globular swelling at the midforehead, lumbosacral myelomeningocele, flexion deformity of the metacarpophalangeal joints of both hands, claw toes, small and deformed ears, and interatrial septal defect (Fig. 12).

The mass at his forehead, which was about one-fourth the size of the child’s head, was soft, cystic, and did not transilluminate. It extended from the nasion to the anterior fontanel. Bone edge with evverting margins was palpable at the base of the mass. The posterior fontanel was closed. The ovoid defect in the skull was bounded on both sides and anteriorly by frontal bones. At its antero-inferior margin, a 1 cm metopic suture was seen extending downward to join the frontonasal suture. Posteriorly the defect merged into the anterior fontanel. The sagittal and coronal sutures were in normal positions. The rest of the skull showed the so-called "beaten-silver" appearance. Both of the frontal lobes extended into and filled the frontal swelling. A constriction ring could be seen on the cerebral cortex corresponding to the rim of the bone defect. The ventricles were slightly enlarged but the brain was otherwise normal.
of the right cyst between the widely separated eyes (Fig. 13).

Upon dissection of the skull, a large defect was found extending from the ethmoidal to the occipital bones. It was bounded by the frontal and parietal bones on both sides. The right orbit was smaller than the left; they were far apart, separated by the rostrally displaced nasal structures. There was a cartilaginous spicule resembling a crista galli between the two nasal openings. Less than half of the brain was within the cranial cavity. The majority of the cerebral cortex was herniated into the left mass, from which a secondary herniation extended into the larger right mass. The left mass was largely filled with brain tissue, while that on the right one was mostly cystic. The portion of the brain tissue in the right mass was partially macerated.

Discussion

Congenital encephalomeningocele is an extracranial protrusion of the cerebral tissue and meninges through a congenital defect in the cranial bones. Classification is unsatisfactory based on the contents of the cranial meningocele, encephalomeningocele, and encephalocystocele, whether they contain pure meninges, meninges and brain, or part of the ventricle, because they can be classified only after a detailed microscopic examination. Classification based on the location of the mass, such as occipital, parietal, vertical, sincipital, basal, nasal, orbital, etc., is useful in the initial description of the patients. In certain areas, however, a single term is used with different meanings, and the different groups are often not clearly defined. The most useful classification appears to be based on the location of the defect in the cranial bones. Here, confusion still exists when one is dealing with the lesion in the anterior and basal part of the head, which is rare in western countries.

According to Mustakallio, in 1855 classified the 19 sincipital hernias of the brain collected by him into nasofrontal, naso-ethmoidal, and naso-orbital subgroups, according to the location of the external orifice of the skull defect. In 1890, von Meyer reported a case and published the same classification, which has since been widely quoted. In 1903, Stadfeldt, quoted by Strandberg, called all three forms of sincipital encephalomeningocele by the general name “fronto-ethmoidal” and gave the following reason: “because it is generally characteristic of all these groups that there is inside the cranium an internal orifice of the hernia distinguishable between the frontal bone and the ethmoid.”

Fig. 11. Case 10. Naso-orbital subgroup of frontoethmoidal encephalomeningocele. Postmortem dissection showing the thick-walled sacs. This patient is Case 11 in our previous report.

Fig. 12. Case 11. Interfrontal encephalomeningocele in an 8-day-old girl who also had a lumbo-sacral myelomeningocele, flexion deformity of the metacarpophalangeal joints of both hands, claw toes, and an interatrial septal defect.
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Fig. 13. Case 12. Anterior-cranial upper-facial cleft. This newborn boy had two large cystic masses overlying a large skull defect. He also had a complete cleft lip and palate on the left side.

Since that time, many reports on the subject have appeared, but the nomenclature and classification are still not uniform. The type characterized by a defect between the frontal and nasal bones with a mass at the glabellar region (our Group 1, Subgroup A) is called nasofrontal by most authors. It has been also called frontonasal, glabellar, nasal, and rhinoencephalocele. The defect in Group 1, Subgroup B, has been called naso-ethmoidal, as well as a more general term such as nasal, anterior, or sincipital. Naso-orbital, anterior orbital, and nasolacrimal have been used for Subgroup C.

The interfrontal type, as in our Case 11 has been reported in a small number of patients. The basal encephalomeningocele is rare and an intranasal mass from herniation through a defect in the cribriform plate is the most common form. Fenger in 1895 quoted Heinecke (1882) as the originator of its classification into sphenopharyngeal, spheno-orbital, and sphenomaxillary, depending on the tract of the herniation. Transtentorial, and intranasal were later added. Blumenfeld in 1965 suggested another classification in accordance with the site where the herniation leaves the cranial cavity, the subgroups being transtentorial, sphenothentorial, trans-sphenoidal, and sphenomaxillary. Most cases of basal encephalomeningocele reported can be included in the former three subgroups. The sphenomaxillary subgroup with a mass in the sphenomaxillary fossa is a theoretical one, and we have found no report of such an occurrence. The encephalomeningocele in the posterior part of the orbit with a defect in the sphenoid wings and orbital plate of the frontal bone cannot, however, be placed in any of the subgroups. It may be called a frontosphenoidal or sphenorostral subgroup.

Patients have been reported with more extensive herniation through a cleft involving the anterior two-thirds of the cranial vault and the upper face, as well as patients with a combination of basal encephalomeningocele and cleft palate, cleft lip, or bifid nose. An extensive occipitocervical defect constitutes the occipital counterpart of the craniofacial cleft. Acrania and anencephaly can be considered to be the most extreme malformations in the same line.

A classification of encephalomeningoceles based on the location of the defect in the cranium is summarized below:

**Classification of Encephalomeningoceles**

I. Occipital encephalomeningocele
   A. Interfrontal
   B. Anterior fontanel
   C. Interparietal
   D. Posterior fontanel
   E. Temporal

II. Encephalomeningocele of the cranial vault
   A. Interfrontal
   B. Anterior fontanel
   C. Interparietal
   D. Posterior fontanel
   E. Temporal

III. Fronto-ethmoidal encephalomeningocele
   A. Nasofrontal
   B. Naso-ethmoidal
   C. Naso-orbital

IV. Basal encephalomeningocele
   A. Transtentorial
   B. Sphen-oethmoidal
   C. Transsphenoidal
   D. Frontosphenoidal or spheno-orbital

V. Cranioschisis
   A. Cranial—upper facial cleft
   B. Basal—lower facial cleft
   C. Occipitocervical cleft
   D. Acrania and anencephaly.

We have found this classification useful in the selection of the operative approach in repairing the lesion. In the nasofrontal subgroup of the fronto-ethmoidal encephalomeningocele, the mass usually presents ex-
tracranially and the lesion has a short neck. This type of lesion can therefore be repaired by an extracranial approach similar to other encephalomeningoceles of the cranial vault. On the other hand, the sacs of the naso-ethmoidal and naso-orbital types have long necks; the distance between the internal and external orifice is considerable, and a good dural closure at the internal orifice is not feasible if approached extracranially. A craniotomy with intracranial repair of the defect is necessary even though it is a more formidable operation; in fact more than one operation may be required. Plastic reconstruction may be done at the same time as the extracranial repair; this is not feasible in the intracranial operation.

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