FRONTO-ETHMOIDAL ENCEPHALOMENINGOCELE

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Congenital malformations are among the most distressing diseases affecting mankind. Many of them are not amenable to reconstructive attempts and must be accepted as a failure of nature. In the majority, the causative mechanism is obscure and prevention is impossible. Geographical variations of various malformations are well known. In Thailand, encephalomeningocele in the front part of the head stands out as an exceptionally common and most interesting malformation of the nervous system.

In the past 7 years, we have treated 82 patients with encephalomeningocele in the front part of the head which may be called sincipital or fronto-ethmoidal encephalomeningocele. The present communication is an analysis of our experiences with these patients at the Chulalongkorn Hospital Medical School, Bangkok, Thailand.

Incidence

Among 21, 071 livebirths in the maternity ward of the Chulalongkorn Hospital during a two and a half year period from June 1963 to November 1965, four fronto-ethmoidal encephalomeningoceles were found. In a survey of a rural community of Nong Ree District, Kanchanaburi Province in Western Thailand[14], a case of encephalomeningocele was found among a population of 6124. The incidence of this disease in Thailand is therefore about 1 in 5000. There are at least one or two patients with this disease in our neurosurgical ward of 33 beds at all time. It comprises about 2 percent of our neurosurgical operations. This occurence appears to be more common than that reported from other neurosurgical centers around the world.

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Etiologic evaluation

All encephalomeningoceles in this series were congenital in origin; the lesions were found at birth. There was no family record of congenital malformation. No two patients in this series were in the same family.

One girl of a pair of twins thought to be identical had a meningocele at the root of nose while the second twin was completely normal.

A study of 26 family trees of these patients up to three generations revealed that all members of 23 families were born in Thailand and were said to be of "Thai blood." In three families the grandfathers or grandmothers on one side were Chinese immigrants; the other half was Thai. None of the patient had Chinese or Indian immigrants as both paternal and maternal ancesters. The significance of this finding is uncertain and needs further evaluation. Considering the big immigrant population of Thailand and the proportion of their admission to our hospital, it might indicate a preponderance of this disease among the Thai population.

Chromosome study was done in 5 patients in this series by our pediatric colleague[3], and all but one were normal. In one newborn, a questionable trisomy was found.

The distribution of blood groups in these patients showed no significant difference from the unaffected population of Thailand.

There was no predilection concerning mother's age and parity. There was no striking history of trauma, infection or metabolic disturbance during the patient's pregnancy.

Pahology. (Figure 1–3)

a. External features. The encephalomeningocele presented as a mass or masses at many sites ranging from the middle part of the forehead, the root of the nose or the side of the base of nose. (Table 1).

The majority of the masses were solid and firm but some were soft and cystic. Transmitted pulsation was detectable in only a small number of patients.

The most common site (32 cases) was in the midline, at the root of the nose, between or just below, the inner canthi. The size of the mass varied from slight elevation to one larger than a child's head. The covering skin varied from thin and shiny or thick and wrinkled. A congenital scar was commonly seen on top of the mass. Hyperpigmentation of skin and hypertrichosis was noted in some patients.

Twenty-three patients had a mass at the glabella or midforehead. The majority of the masses were sessile but a few were pedunculated. Three patients with mass at this location were born with exposed brain tissue without skin covering. Continuous bleeding and dripping of cerebrospinal fluid was noted in these patients. The rest had skin covering of variable thickness.

Table 1. Site of mass of encephalomeningocele

Glabella	23
Middle part of root of nose between the eyes	32
One side of base of nose	10
Both sides of base of nose	6
Inside the nose (widened base)	6
Lower part of bridge of nose	2
Faceal cleft	2
Multiple sites	1
	82

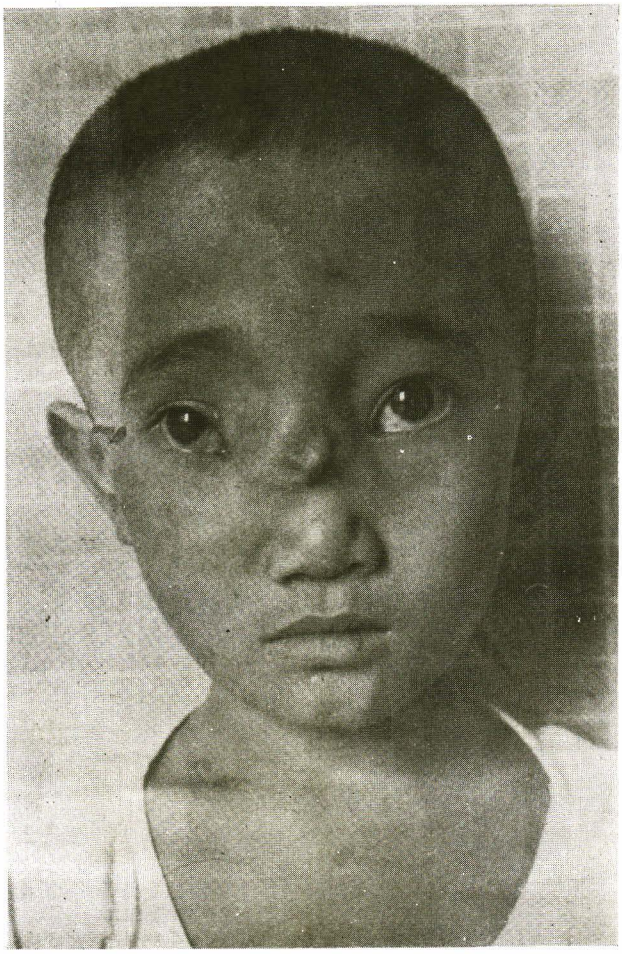

Figure 1. Seven-year-old boy with a mass about the size of a finger tip at birth and progressively enlarging. Note the scar-like marking on top of the mass.

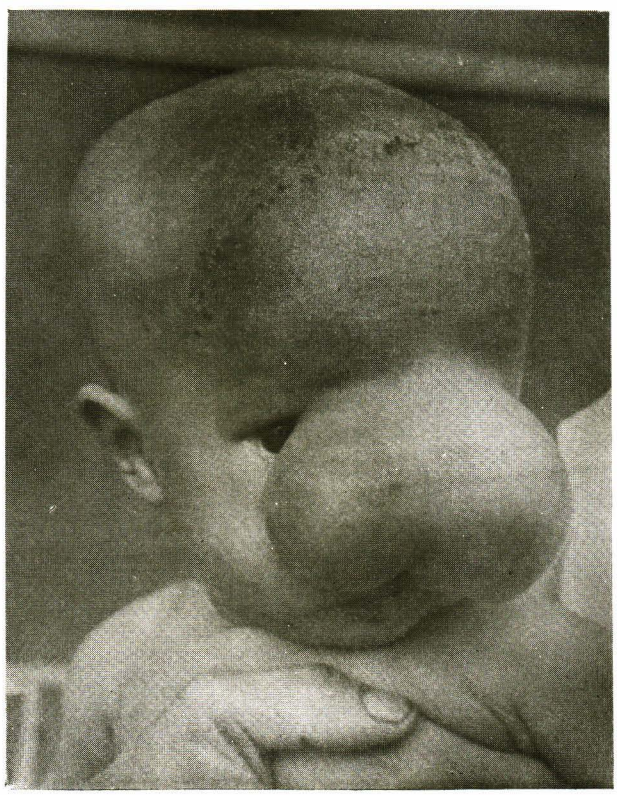

Figure 2. A 3-month-old girl with a big mass at the glabella. Her respiration was interfered by the mass when she lay in supine position.

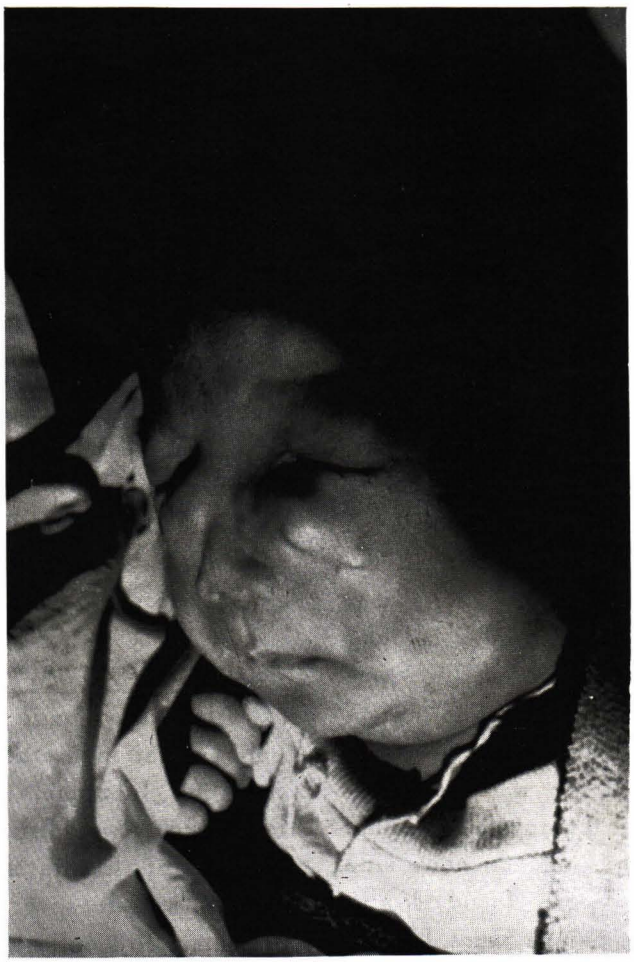

Figure 3. A 1—month—old boy with firm masses at both sides of his nose.

Mass at the lower part of nose and on one side of the base of the nose were less commonly seen. The character of the mass and covering skin was similar to the ones described.

In 6 patients the mass was inside the nasal cavity and only widening and swelling of the root of the nose was seen externally. The nasal bone overlying the mass was expanded but intact. The covering skin was normal.

Deformity of the face with microcephaly and facial cleft in addition to the huge encephalomeningocele was encountered in two patients.

b. Skull defects (Fig. 4–7)

The defect of the cranial bones through which the herniation passed was examined at operation in 59 patients. In all, the opening was located between frontal and ethmoidal bones. One opening in midline was found in 30 patients. The crista galli was identified at its posterior rim. The site of the defect corresponded to that of the foramen cecum. Bilateral openings, one on each side of the midline, were found in 18 patients. The openings were located at the anterior end of the cribriform plates and the crista galli was at the posterior part of the bridge of bone between them. A unilateral opening at the anterior end of cribriform plate was seen in the remaining patients.

Postmortem dissection was done in 5 patients. The location of the cranial end of the defect, being at the junction between frontal and ethmoidal bone, was confirmed. The location of the overlying facial bones, namely, nasal, lacrimal and maxillary bones was variable. In some, the nasal bone attached to the upper edge of the maxillary bone at the inferior aspect of the mass. In such cases, the mass was high at the glabella or midforehead.

In the other cases, the nasal bone was at the lower edge of the frontal bone above the mass of the encephalomeningocele. In one patient the facial bones were in normal positions and the encephalomeningocele had an elongated neck situating between the nasal and lacrimal bones.

In addition to the defect, there was usually hypertelorism and an increase in the depth of the anterior cranial fossa.

c. Brain abnormalities (Fig. 8)

All but two patients operated upon had herniation of the frontal tips into the sac. In some, the intervening falx cerebri was also protruding. The amount of herniated brain varied from a half to several centimeters in diameter. In small number of patients, the anterior horn of the lateral ventricle was also involved.

When large amount of brain tissue was herniated, the head was consequently small. The whole brain was pulled foreward and the optic nerves, internal carotid arteries and pituitaty stalk were angulated anteriorly. The tip of the temporal lobe was seen to herniate above the

Figure 4. Postmortem dissection of the child in figure 3 showing markers in the defects between lacrimal and frontal process of maxillary bones.

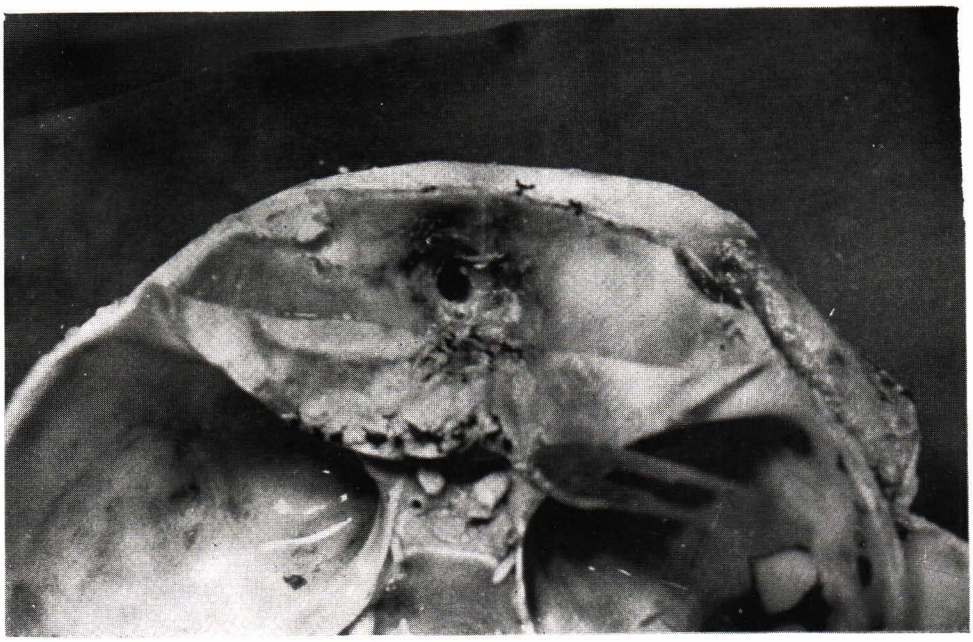

Figure 5. Floor of the cranial cavity of the child in fingure 3 showing defects at the anterior end of the cribriform plates. The crista galli was at the posterior part of the bridge of bone between the two openings.

Figure 6. A newborn with a mass of exposed brain tissure at the glabella. The head was small and the fontanels were closed. The mass bled easily and the child expired from blood loss.

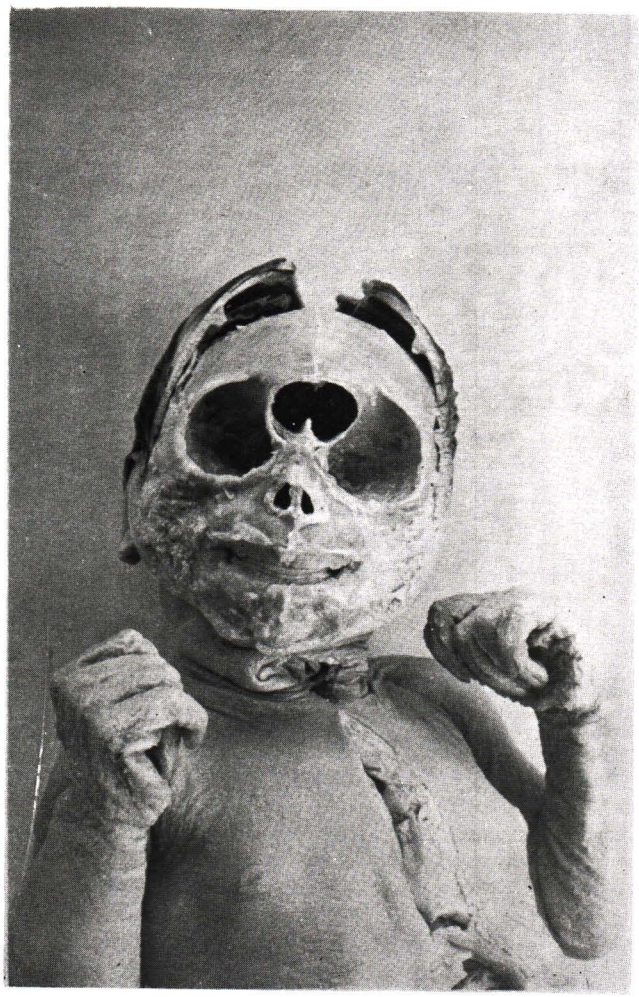

Figure 7. Postmortem dissection of the child in figure 6 showing a large defect in midline corresponding to the foramen cecum. The crista galli was identified at its postero—inferior rim. The nasal bones were below the defect.

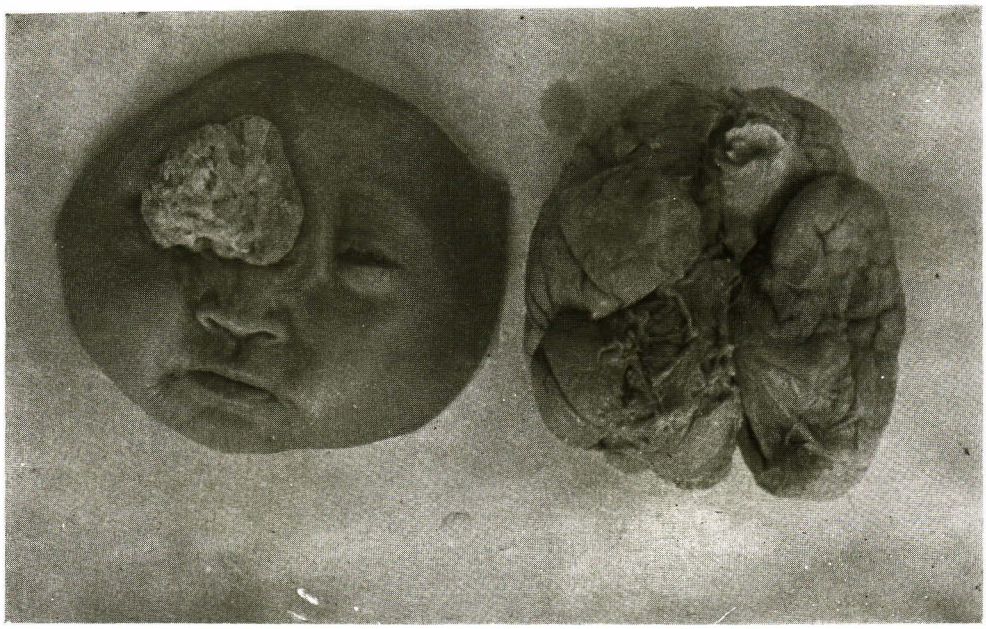

Figure 8. Brain removed at autopsy from a 1—month old child with a mass at the right side of glabella. Herniation of the tip of left frontal lobe was associated with left unilateral hydrocephalus. Obstruction of left foramen of Monro was due to the deformation of brain secondary to the herniation.

sphenoid ridge into the anterior cranial fossa in one patient. The foramen of Monro might be partially or completely occluded resulting in unilateral or bilateral hydrocephalus.

Holoprosencephaly, agenesis of corpus callosum, localized cortical atroply were sometimes seen in association with the encephalomeningocele.

d. Eye abnormalities

Hypoplasia of one or both eyes was also seen associated with the brain abnormalities. The palpebral fissure, orbit and eyeball of the affected eye was small and the pupillary opening was absent.

Clinical features

These encephalomeningoceles presented at birth but the patients came to us at ages ranging from premature newborn to 25 years. At birth the lesion was usually smaller, some of them showed only a small scar at the root of the nose and hypertelorism. As the child grew, the mass progressively enlarged and the facial deformity increased.

Generally the lesion was quite apparent and there was no problem in the diagnosis of the classical cases. This could, however, be difficult in patients where the opening was small and the communication between the sac and the intracranial cavity was not readily detectable. The lesion had to be differentiated from tumors or cysts indigenous to the region.

Aspiration of the mass and determination of the sugar content of the fluid was helpful in some patients. Plain roentgenography of the skull was very helpful, the diagnosis of the encephalomeningocele being suggested by the abnormally wide distance between the two orbits. As a rule, the opening could not be seen in a newborn infant due to poor calcification of the cranium, but in older children it was usually visible. Postero–anterior projections at various angles was helpful because the opening was best seen only at certain angles. Occasionally tomography was helpful in defining the opening more clearly. Scanning of the head after intrathecal injection of radioactive iodinized serum albumin was helpful in demonstrating the communication between the sac and the intracranial cavity. In the past 3 years, pneumoencephalography was used routinely in order to outline the neck of the sac as well as to find the associated abnormalities of the brain. (Fig. 9)

Treatment.

a. Indication

The bizarre nature of the encephalomeningocele in the middle of the face makes removal almost mandatory for cosmetic reason alone. However, in some patients, the swelling progressed with age and surgical correction obviates the brain damage

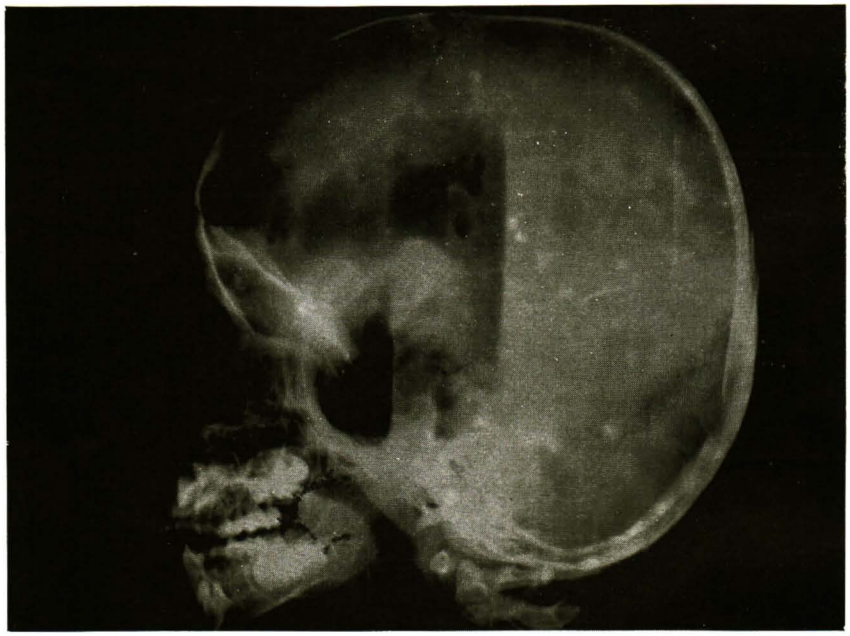

Figure 9 Pneumoencephalogram in a boy with encephalomeningocele showing ventricular dilatation and cortical porencephalic cysts.

caused by further herniation of brain tissue into the sac. Furthermore, ulceration, rupture and leakage of cerebrospinal fluid is probable and removal will prevent subsequent meningitis. Finally removal of a big mass facilitates care of the infant.

b. Contraindication

Not every patient should be submitted to surgery. In our opinion, it is not justified in children with severe brain damage or little hope of normal mental development. Massive herniation of tissue, associated brain anomalies and hydrocephalus are the causes of brain damage. In the presence of acute infection of the ulcerated sac, the operation should be postponed.

c. Optimal time for surgical repair

In patients born with a mass of exposed brain tissue protruding from the middle of the forehead, an emergency operation is necessary in order to stop the continuous bleeding and to prevent the inevitable infection. In general, surgical repair should be carried out just as early as the child's condition permits. Early surgery will minimize the facial deformity and other complications. It will also prevent brain damage from further herniation into the defect.

d. Operative techniques

A number of patients were seen with complications resulting from simple excision, done at other hospitals. Leakage of cerebrospinal fluid through disruption of the wound was not uncommon. The patients had recurrence of the mass with additional brain damage. Meningitis also increased the likelihood of development of hydrocephalus.

In our experience a successful treatment requires a good water-sealed closure of the intracranial end of the herniation. If the hole in the bone is of considerable size, a strengthening of the bone defect is also essential. We have elected to use a fascial or periosteal graft to repair the dura and a stainless steel mesh extradurally over the defect.

A good repair can be accomplished by two approaches, an extracranial and an intracranial one.

1. Extracranial approach

An elliptical incision is made around the base of the herniated mass. Care is taken to have enough skin for closure of the wound. Amputation of the sac is at the level of the skull defect. The herniated brain is removed. A piece of periosteum or temporal fascia is then sutured to the dura with interrupted silk sutures at 1–2 millimeter intervals. A piece of stainless steel mesh is then inserted across the bone defect between the dura and skull. Galeal and subcutaneous sutures are applied before a fine skin closure.

2. Intracranial approach

Through a coronal incision a bilateral frontal craniotomy is done,

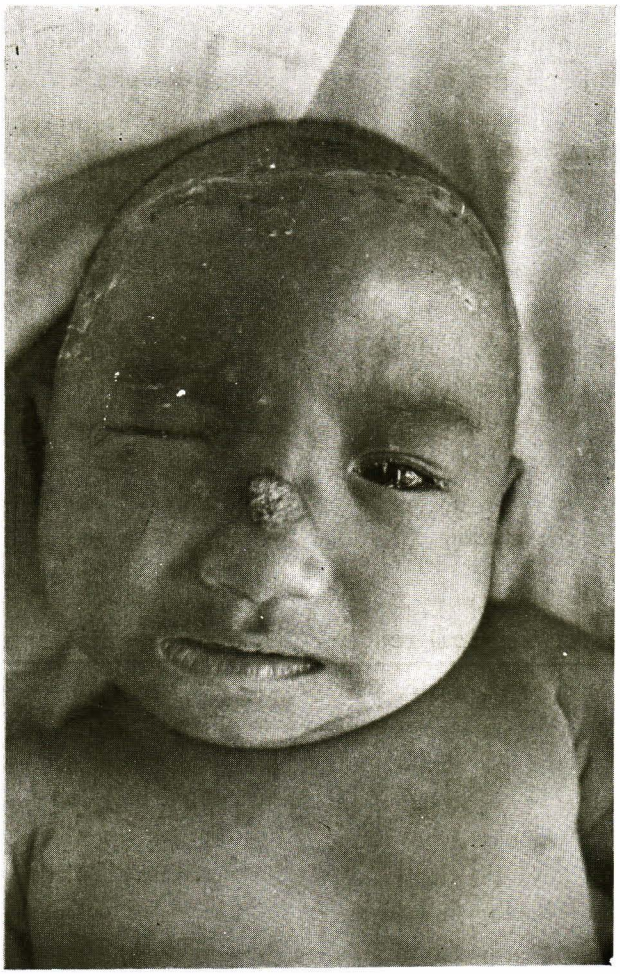

Figure 10. Postoperative photograph of a 2 month old girl showing the collapsed sac of encephalomenigocele at root of the nose. Note wide intercanthal distance and hypoplastic right eye.

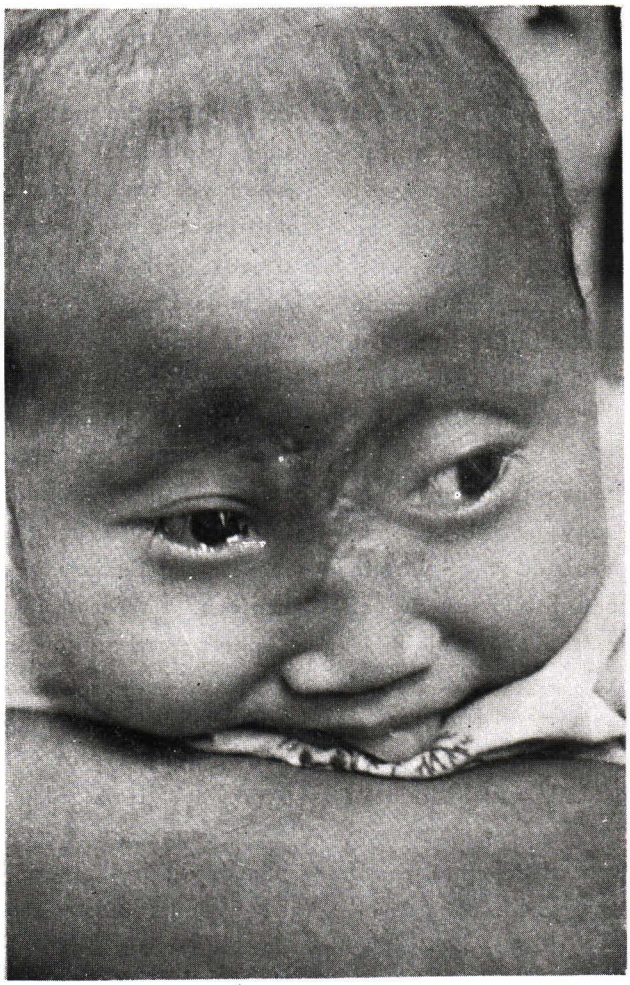

Figure 11. Postoperative photograph after a secondary V—Y plastic repair which was done one month after the intracranial operation.

Exploration is made under the frontal lobe both inside and outside the dura. The herniated brain is amputated at the cranial end of the orifice and the herniated portion is partially removed. The dura is elevated from the bone around the rim of the opening. Some difficulties may be encountered at the posterior edge where the crista galli is situated. A piece of periosteum or fascia is then sutured to the dural defect. A very small curved needle may be needed in making the deepest sutures. The bone defect is then reinforced by extradural stainless steel mesh.

After the intracranial operation, the facial mass may be obliterated. In some patients, a second stage plastic operation is needed for removal of the excessive skin and the remainder of the sac as well as correction of ocular hypertelorism and flatness of the nasal bridge. This can best be effected by using a V–Y technique. (Fig. 10, 11)

Advantages and disadvantages of each approach have to be considered in selecting the operative method. The extracranial operation is feasible only when the lesion has a short canal and the intracranial end of the defect is accessible through the hole. It is therefore suitable for patients with a mass at the forehead or upper part of the root of the nose.

On the other hand, the intracranial approach, being a craniotomy, is a more formidable operation. It is however, necessary whenever the distance between the internal and external orifices is considerable and a good dural closure at the internal orifice is not feasible otherwise.

In a number of patients, hydrocephalus is the more pressing problem and a shunting operation is done whenever the ventriculogram shows sufficient cortical preservation. Operative repair of the encephalomeningocele is deferred.

Discussion :

Ingraham and Matson [5] found reports of 87 encephaloceles as compared to 1157 spinal lesions. Of the encephaloceles, only 21 cases were in nasal, nasopharyngeal and frontal regions. There have been occasional reports of meningoceles in the anterior aspects of the head and in the nose (2,4,6,7,8,9,16) but the condition is considered a rarity in Europe and America. It is also true in Japan, Hongkong and Southern India. In Thailand, it is exceptionally common; 82 patients being seen in 7 years at our neurosurgical service. High incidence are also found in Malaysia and Indonesia. Tandom [15] reported a higher than average incidence at Lucnau, Northen India but stated that the incidence in New Delhi was low. For the African Negro in Nigeria, Odeku [10] found 6 sincipital ones among 36 encephalomeningoceles. He also quoted Gupta who found one encephalomeningocele among 4220 births

at a hospital in Ibadan, Western Nigeria, Acquaviva reported a big series of 39 patients with sineipital encephalomeningocele in Morrocco. The incidence in Africa is therefore slightly higher than in Europe and America but is still lower than in Southeast Asia.

Meningoceles situated in the anterior part of the skull are sometimes referred to as sincipital encephalomeningoceles and are usually classified according to the site of the protruding mass. The findings at operation or autopsy in our cases, however showed that the persistent site of the cranial opening in all cases was at the junction between the frontal and ethmoidal bones. In the majority the opening was in the midline and the site corresponded to that of the foramen cecum or anterior neuropore. The site of the protruding mass varied according to the development of the nasal, lacrimal and frontal processes of maxillary bones overlying the defect. That the site of the cranial opening in our patients was so often the same suggests common etiologic factors. Consequently we believe that these encephalomeningocele can be identified as a fronto–ethmoidal type.

Summary :

We have studied 82 patients with encephalomeningocele in the anterior part of the head. The geographical variations in the incidence of this disease have been discussed. Pathological, clinical and therapeutic aspects have also been considered.

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