Fronto-ethmoidal Encephalomeningocele

TAKASHI HAYASHI, AKIRA ISHIBASHI, MASAHIKO KATAYAMA, TAKEO HASHIMOTO*, JIRO NISHIMURA**, EIICHIRO HONDA† AND SHINKEN KURAMOTO†

Departments of Neurosurgery, Neonatology* and Plastic Surgery**, ST. Mary's Hospital and †Department of Neurosurgery, Kurume University School of Medicine, Kurume, 830 Japan

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Summary: A male child, born by normal spontaneous vaginal delivery, was noted to have a soft tumor, in the shape of an "elephant's nose", with a diameter of 2.5 cm × 2.5 cm and 5.5 cm long in the inter-orbital region. With the diagnosis of naso-frontal type of fronto-ethmoidal encephalomeningocele with congenital hydrocephalus, corrective surgery was performed in two stages. At the first operation, a ventriculoperitoneal shunt and bilateral frontal craniectomy were performed, and closed with frozen human dura mater intracranially. Twenty days after the first operation, the encephalomeningocele was excised. The patient tolerated the surgery well.

Key words: cranium bifidum — frontal encephalocele — dysraphic states — congenital hydrocephalus — craniolacunia — encephalomenigocele — macrocrania

Introduction

Among the different forms of cranium bifidum, cranium bifidum occipitalis is the most common. In contrast, cranium bifidum frontalis is very rare. In this paper we describe a case of fronto-ethmoidal encephalomeningocele associated with congenital hydrocephalus.

Case Report

The patient is a male child born by normal spontaneous vaginal delivery. At birth, a soft tumor, in the shape of an "elephant's nose", with a diameter of 2.5 cm × 2.5 cm and 5.5 cm long was noted in the inter-orbital region (Fig. 1). The tumor was noted to expand with crying; it was consequently suspected that the tumor communicated with the cerebrospinal fluid pathway.

The head circumference was 35.5 cm and within normal range; the baby's length was 49 cm, and the weight was 3,080 gm. On
neurological examination, there was good movement of the limbs, with normal sucking reflex, Moro reflex and traction reflex.

**Neurological findings:** Minimal craniofacial disproportion and craniolacunia were seen on the lateral view of the plain x-ray films of the skull. A bony defect was noted to extend to the fronto-nasal junction from the glabella (Fig. 2). On CT scan a dilatation of the cerebral ventricles was noted (Fig. 3-a). On the CT scan 1 cm above the ethmoidal plate, a bony defect at the naso-frontal junction was noted. A part of the frontal pole extended out of the cranium through this defect (Fig. 3-b).

In light of these findings, we diagnosed the patient with fronto-ethmoidal encephalomeningocele of the naso-frontal type, according to the classification of Suwanwela and Suwanwela (1972).

**Surgical treatment:** The surgical man-

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**Fig. 2.** Lateral view of the x-ray film of the skull shows minimal craniofacial disproportion and craniolacunia. A bony defect is noted to extend to the fronto-nasal junction from the glabella.

**Fig. 3-a.** CT scan shows ventriculomegaly indicating congenital hydrocephalus. Arrow shows ventricular catheter of V-P shunt.

**Fig. 3-b.** Bone-window CT image at 1 cm above the ethmoidal plate shows a bony defect at the naso-frontal junction and a part of the frontal lobe extended out of the cranium through the defect.
management of this case was divided into two parts. The first operation was carried out on the 7th day after birth, with the installation of a V-P shunt, followed by bilateral craniectomy with a coronal incision of the scalp. When the frontal lobes were exposed, they were seen to herniate outside of the cranium through the bony defect (mainly on the left side, but also partially on the right side). The brain matter was covered with dura mater. These portions of the brain were resulted to the cranium as much as possible, and the portions of the brain which were discolored and necrotic were excised. The defect in the dura mater was corrected as much as possible with a crumpling stitch. We then secured human frozen dura (Lyodura) to the suture-exposed dura mater at its edges, thus completely correcting the dural defect (Fig. 4-a, b). On the 23rd day following the first operation (the 30th day after birth), the tumor on the outside of the cranium was excised. Histological examination of the contents

Fig. 4-a. Herniated frontal lobes are seen at operation.

Fig. 4-b. Schematic drawing of the first stage operation.
   a: herniated frontal lobes, b: fronto-nasal bony deficit, c: covering over the bony deficit with a human dura mater (Lyodura).
revealed that the tumor consisted of glial tissue.

Postoperative course: The patient did well following both procedures, and left the hospital 10 days following the second operation (i.e., 40 days following birth). Neurological examination at the time of discharge from the hospital showed no abnormal findings with respect to movement of the limbs. Figure five is a photograph of the baby taken at the time of discharge.

Discussion

Matson (1969) predicted that frontal encephalocele would rarely occur among individuals of European stock, but that it would occur relatively more frequently among Asian. In support of this hypothesis, Suwanwela (1971) presented 100 cases of cephalocele frontalis (sincipital encephalomeningocele) which they observed during 8 years in Thailand, and applied their classification to this series. However, Flatz and Sukutomiya (1970), in their investigation of the frequency of cephalocele frontalis in Northern Thailand, were unable to determine any hereditary or environmental factors. In Japan, Akashi et al. (1969) analyzed 33 cases of cephalocele which they had encountered and reported that 29 cases out of 33 (87.9%) were cephalocele occipitalis. These findings do not support the conclusion that cephalocele frontalis is particularly common among Asian. With respect to the classification of cephalocele frontalis, Davis and Alexander (1969) reported on 7 cases which they experienced into three groups according to the classification of Davis and Alexander (1969): fronto-ethmoidal encephalomeningocele (10 cases); inter-frontal encephalomeningocele (1 case); and cranio-facial cleft (1 case). The first category includes three subtypes: naso-frontal (6 cases), naso-ethmoidal (3 cases) and nasoorbital (1 case). Among the fronto-ethmoidal encephalomeningoceles, the naso-frontal type occurs most frequently. Our own case is equivalent to the naso-frontal type as classified by Suwanwela and Suwanwela (1972).

In treating the patient reported here, we followed the approach of Matson (1969) in returning the herniated brain to the inside of the cranium as much as possible, operating intracranially in order to avoid leakage of cerebrospinal fluid. We carried out a V-P shunt for the congenital hydrocephalus as the initial stage, returning the prolapsed brain through a bifrontal craniectomy and correcting the dural defect. This was followed by a second operation involving excision of the tumor directly from the outside of the cranium.
References


