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Meningomyelocele associated with cranium bifidum: rare coexistence of two major malformations

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Abstract. Three cases of lumbosacral meningomyelocele associated with cranium bifidum are reported. The meningomyeloceles were of the neural placode type and the cranium bifidum was an interfrontal encephalomeningocele in all cases. Two of the patients received ventriculoperitoneal shunts and had evidence of type II Chiari malformation on magnetic resonance imaging. No evidence of a Chiari malformation was revealed in the third patient. We discuss the etiology of the rare coexistence of these two major malformations.

Key words: Meningomyelocele – Cranium bifidum – Interfrontal encephalomeningocele – Chiari malformation

Introduction

Open neural tube defects of the spinal cord occur at an incidence of 1.0–1.5 per 1000 live births in North America and 0.2 per 1000 in Japan [3], while the worldwide incidence of cranium bifidum is approximately 1 per 5000 live births [5]. Both of these entities are believed to be related to defective closure of the primitive neural tube, a phenomenon that is underway or complete by the 6th week of intrauterine life. However, there have been no reports of patients who have both meningomyelocele and cranium bifidum, apart from one case described by Suwanwela and Suwanwela in 1972 [6]. Here we present three cases in which lumbosacral meningomyelocele was associated with cranium bifidum of the interfrontal type and describe their clinical course.

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Case reports

Case 1

The patient was a 9-year-old boy. He had been delivered spontaneously after a 41-week gestation to healthy parents. His birth weight was 3200 g, head circumference was 39 cm, and Apgar score was 9. His anterior fontanel was tense, and fluctuant mass the size of a fist and covered with normal skin was noted in the midline of the forehead. In addition, lumbosacral rachischisis and pes equinovarus were noted (Fig. 1a,b). There were no other associated anomalies. Neurological examination demonstrated paraplegia and absence of the anal reflex. The anteroposterior skull X-ray film showed an ovoid bony defect encircled by a hyperostotic rim in the midline of the frontal bone (Fig. 1c). Computed tomographic (CT) scans showed marked ventriculomegaly and prolapse of brain tissue including the frontal horn of the lateral ventricle out of the skull through the bone defect. There was an epidural hematoma and a subgaleal hematoma in the left temporal region (Fig. 1d). On the 2nd day after birth, repair of the rachischisis, installation of a ventriculoperitoneal (VP) shunt, and evacuation of the epidural hematoma were performed. At operation, communication between the epidural and subgaleal hematomas via a bone defect in an area of craniolacunia was noted. The postoperative course was uneventful. When the infant was aged 10 months, the frontal bone defect was repaired using autogenous bone fragments (Fig. 1e). Magnetic resonance imaging (MRI) at the age of 9 years revealed the characteristic features of Chiari malformation (Fig. 1f). The child has slight psychomotor delay and uses a wheelchair.

Case 2

This patient was an 8-year-old boy. He was delivered to healthy parents by cesarean section for cephalic presentation after 32 weeks of gestation. His birth weight was 3080 g, head circumference was 41.5 cm, and Apgar score was 9 at 1 min. A rachischisis in the lumbosacral region and a soft fist-sized mass on the forehead covered by normal skin were noted (Fig. 2a, b). Skull X-ray films revealed a bone defect with a hyperostotic rim in the frontal bone and craniolacunia in the frontal and parietal regions. CT scans showed a frontal bone defect and marked ventriculomegaly (Fig. 2c). Spinal X-ray films demonstrated spina bifida from L2 to L5. On the 2nd day of life, repair of the meningomyelocele was performed, followed by insertion of VP shunt. Although cranioplasty for the frontal bone defect was scheduled, the parents refused further treatment. MRI at 8 years of age revealed the Chiari malfor-

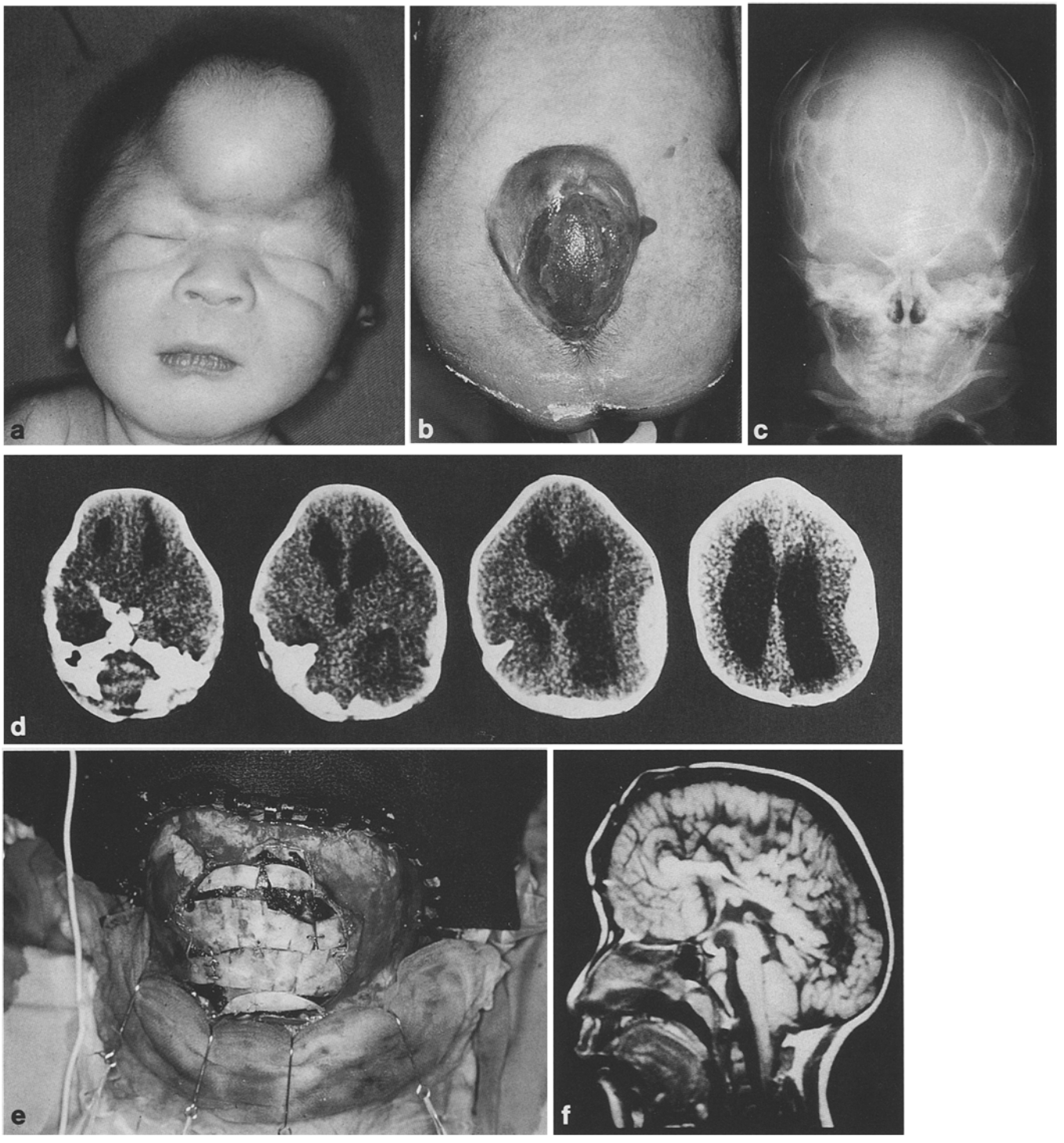

Fig. 1a–f. Case 1. **a, b** Photographs taken at birth show **a** an interfrontal encephalomeningocele and **b** lumbosacral meningomyelocele. Note the typical appearance of the neural plaque. **c** Anteroposterior view of skull X-ray film shows an ovoid bony defect encircled by a hyperostotic rim in the midline of the frontal bone. **d** Computerized tomography (CT) shows the epidural hematoma and marked ventriculomegaly. **e** Intraoperative photograph during the repair of the frontal bony defect. The frontal bony defect is repaired using autogenous bone fragments. **f** Magnetic resonance imaging (MRI) at the age of 9 years shows the characteristic features of Chiari malformation. Note the frontal lobe without developmental failure

mation (Fig. 2d). The boy now has moderate psychomotor delay and uses a wheelchair.

Case 3

The third patient was a 4-year-old boy. He was delivered spontaneously after 39 weeks of gestation. His birth weight was 3150 g, head circumference was 34.5 cm, and Apgar score was 10. A soft bulge covered by normal skin was associated with a bone defect of the forehead and protruded anteriorly, becoming tense when the child cried. A rachischisis partially covered by normal skin was also

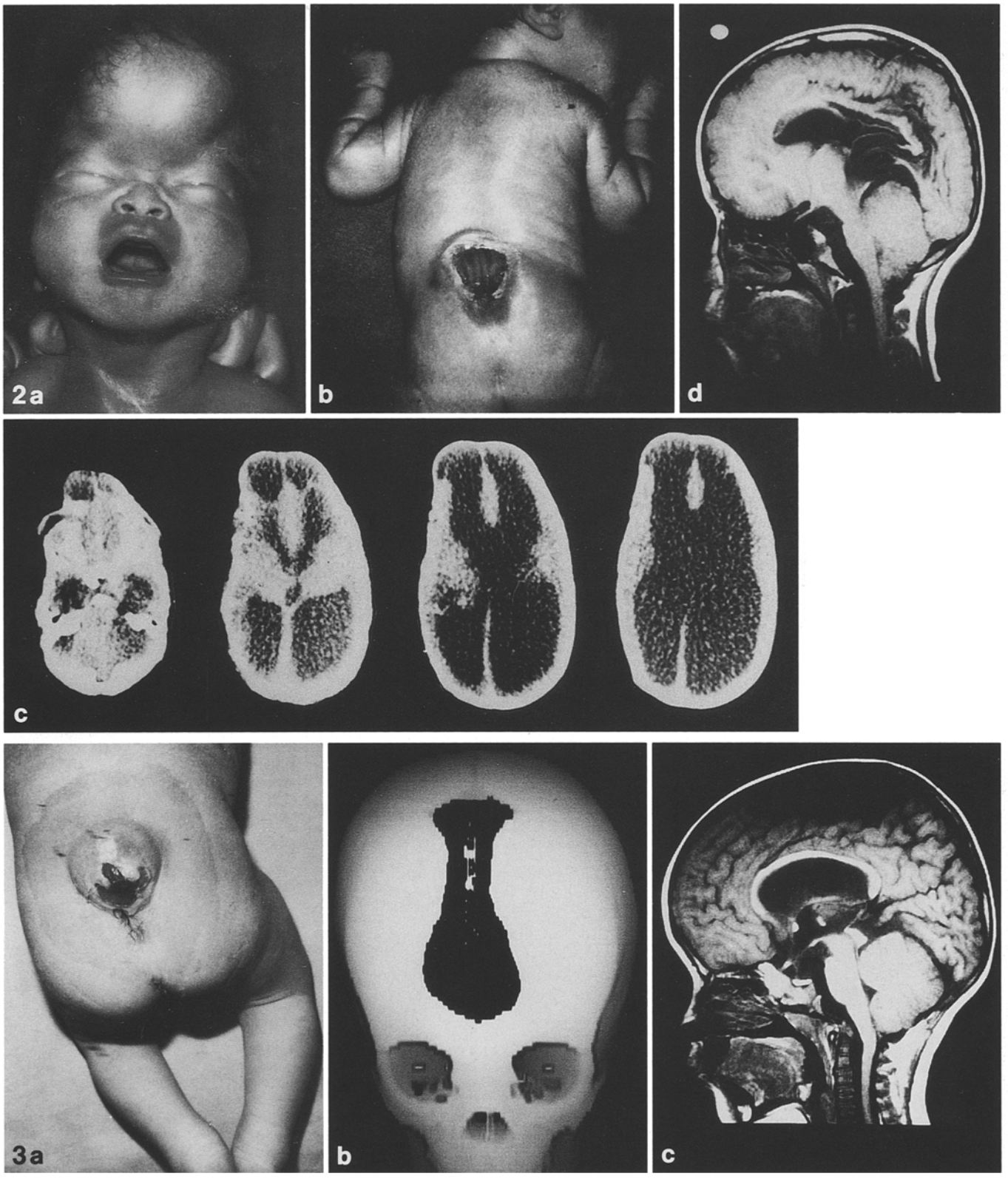

Fig. 2a–d. Case 2. **a, b** Photographs taken at birth show an interfrontal encephalomeningocele (**a**) and lumbosacral meningomyelocele (**b**). **c** CT shows marked ventriculomegaly. **d** MRI at 8 years of age shows characteristics of the Chiari malformation. Note the intact frontal lobe

Fig. 3a–c. Case 3. **a** Photograph taken at birth shows lumbosacral meningomyelocele partially covered by normal skin. **b** Three-dimensional CT display of the skull showing a bony defect in the frontal bone. **c** MRI at the age of 4 years shows normal development

found in the lumbosacral region (Fig. 3a). Neurological examination including lower limb movements and the anal reflex revealed no abnormalities. A bone defect in the forehead without evidence of craniolacunia was demonstrated by plain skull films. CT scans showed slight enlargement of the ventricular system. Three-dimensional CT display using the bone algorithm revealed an ovoid defect in the frontal bone that merged into the anterior fontanel (Fig. 3b). Repair of the meningomyelocele was performed on the 1st day of life. The postoperative and subsequent clinical course was uneventful. As repeated CT scans failed to demonstrate gradual enlargement of the ventricles in follow-up studies, VP shunting was not performed. Radiological examinations showed evidence of ingrowth of the frontal bone from the rim of the defect, so repair of the defect in the frontal bone was not performed either. MRI at the age of 4 years failed to demonstrate any evidence of the Chiari malformation (Fig. 3c). The child's neurological and intellectual development have been excellent, without any evidence of neurological deficits.

Discussion

An encephalocele is defined as a protrusion of the cranial contents beyond the normal confines of the skull. This cerebral herniation passes through a congenital cranial defect and may contain brain tissue and/or a portion of the ventricles surrounded by meninges. Suwanwela and Suwanwela [6] divided encephalomeningoceles in the anterior part of the head into the following three groups based upon the location of the bone defect: (1) frontoethmoidal encephalomeningocele, including nasofrontal, nasoethmoidal, and naso-orbital types, (2) interfrontal encephalomeningocele, and (3) craniofacial cleft. In their patient, an interfrontal encephalocele in the middle of the forehead arose through a bony defect in the frontal bones in the region of the metopic suture and the frontal bone inferior to the bony defect bordered the fontanel superiorly. Their patient was identical to our three in the location of the bone defect as well as in the associated lumbosacral meningomyelocele.

No one developmental theory adequately explains all the anatomical variations known collectively as encephalocele. Failure of the anterior neuropore to close at the cephalic end of the neural tube at the level of the foramen cecum in the frontal bone by approximately 24 days of development has generally been accepted as the etiology [2, 4]. However, the secondary herniation of normally developed cerebral tissues shown by MRI indicates that direct comparison with spina bifida aperta is inadequate to provide an explanation. If the neural tube failed to close at this early stage, the cerebral hernia would be expected to contain tissues of the tectum but not of the cerebral hemispheres. Especially in the case of lateral encephaloceles, it has been suggested that the brain protrudes through a defect caused by the failed closure of sutures that routinely close before birth, and this is suspected to occur as a result of raised intracranial pressure due to the associated hydrocephalus [7]. This would be plausible except that there was no evidence of hydrocephalus in one of our three cases. In addition, the rim of the ovoid bone defect was shown to be hyperostotic in our patients, indicating that it was congenital. The traditional theory also fails to explain why more cases with hydrocephalus do not have an interfrontal encephalocele that protrudes through the the anterior fontanel, which is the largest defect in the skull at birth. Hendrick [2] suggested that failure of the development of overlying mesenchymal tissue associated with local cerebral abruption occurring at 8–12 weeks of gestation would cause this anomaly, rather than local failure of the neural tube.

Almost all infants with meningomyelocele have an associated type II Chiari malformation [1, 8]. In two of our three cases, typical features of the Chiari malformation were also observed. In patients with encephalomeningocele, the coexistence of a Chiari type II malformation has not been reported previously. Our cases suggest that the type II Chiari malformation developed in association with the meningomyelocele, because the two patients with this malformation had more typical rachischisis and more severe neurological deficits such as paraplegia and mental retardation.

Conclusion

Three cases of interfrontal encephalocele associated with meningomyelocele were reported. The causes of these two major malformations were suspected to be different. In the interfrontal encephalocele, the normal growth of the cerebral hemispheres suggests that failure of development of the overlying mesenchymal tissue may allow local cerebral abruption to occur, rather than that this condition arises as a neural tube abnormality.

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