Acquired Lacunar Skull Deformity Following Ventriculoperitoneal Shunt Placement
—Case Report—

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Abstract
A female infant developed hydrocephalus as a result of intraventricular hemorrhage related to premature birth. Radiography showed no sign of lacunar skull deformity (LSD). Lateral and fourth ventricle ventriculoperitoneal shunts were placed. The fourth ventricle shunt required replacement at age 3 years. Radiography and computed tomography showed LSD. Follow-up radiography at age 8 years showed the LSD was becoming unclear. LSD generally appears before birth, and disappears shortly after birth. LSD is most commonly associated with spina bifida cystica or encephalocele. This case of acquired LSD indicates that abnormal disorganized collection of collagen fibers of the skull plates may be triggered by iatrogenic intracranial hypotension even after infancy.

Key words: lacunar skull deformity, hydrocephalus, skull radiography, ventriculoperitoneal shunt

Introduction
Lacunar skull deformity (LSD) is characterized by shallow depressions or deep cavitations in the cranial vault. LSD develops before birth and has been demonstrated in utero in the last trimester. LSD is most commonly associated with spina bifida cystica or encephalocele. Here we describe an infant case of acquired LSD which appeared following ventriculoperitoneal (VP) shunt placement.

Case Report
A female infant developed hydrocephalus as a result of intraventricular hemorrhage related to premature birth. Radiography showed no sign of LSD (Fig. 1). Shortly after birth, a VP shunt was placed in the lateral ventricle. An additional VP shunt was placed for trapped fourth ventricle at 8 months after birth, but shunt infection was triggered, so the entire shunt system was removed, and ventricular drainage was undertaken. The infection improved by age 1 year, and VP shunts were simultaneously placed in the lateral and fourth ventricles. Both shunts employed the Codman Hakim programmable valve (Medos S.A., Le Locle, Switzerland), with opening pressures at 60 mmH₂O for both lateral and fourth ventricles. On the 3rd day, the fourth ventricle had expanded and mild impairment of consciousness was observed, so we reduced the opening pressure of the fourth ventricle shunt to 30 mmH₂O.

Fig. 1 Radiograph at birth, lateral view, showing no lacunar skull deformity.
Fig. 2  Radiological studies at the age of 5 years 5 months. A: Radiograph showing multiple lucent defects in the parietal region, compatible with lacunar skull deformity but without digital markings. B: Computed tomography scan showing the honeycomb pattern of the calvarium with multiple areas of bone thinning.

Fig. 3  Radiograph at the age of 8 years showing the lacunar skull deformity becoming unclear.

Mild impairment of consciousness and nystagmus were observed at the age of 2 years 8 months. The lateral ventricle was exhibiting a tendency to enlarge and the fourth ventricle showed a tendency to shrink. Therefore, the opening pressure of the lateral ventricle shunt was reset to 30 mmH₂O, and that of the fourth ventricle shunt was reset to 80 mmH₂O. These adjustments improved the symptoms, but the fourth ventricle shunt failed 4 months after the resetting, so another VP shunt was placed in the fourth ventricle using the same opening pressure. Radiography showed indications of LSD at the age of 3 years 4 months. Twenty-five months after the final resetting of opening pressure, radiography and computed tomography clearly demonstrated LSD (Fig. 2). Radiography showed the LSD becoming unclear at the age of 8 years (Fig. 3).

**Discussion**

Skull radiography shows LSD as a honeycomb pattern consisting of numerous circular or oval lucent defects with a clear border,⁶ which occur in the parietal, frontal, and occipital bones, in descending order of frequency. The honeycomb pattern tends to converge towards the thick portion of the skull along the sagittal, coronal, and lambdoid sutures,²,⁷ and this distribution can also be observed by neurosonography.¹

LSD is probably not associated with increased intracranial pressure. Many infants with LSD present with microcephalus or normal head circumference.⁵ Increased intracranial pressure is not always present. In addition, autopsy studies reveal differences in the tracks of the LSD and the gyral patterns of the cerebral cortex,⁸ which indicate that increased intracranial pressure does not contribute to the occurrence of LSD.

Absence of cerebral ventricular distension caused by decompression of an open neural tube may be the underlying etiologic factor in the development of LSD.⁴ In the absence of distension of the underlying fetal brain, the induction of normal development of the membranous plates of the fetal calvarium cannot take place, resulting in abnormal disorganized collections of collagen fibers, which on completion of ossification cause the characteristic radiographic findings of LSD. Hydrocephalus and/or the normal brain expansion process remodels the skull plates late in gestation or postnatally, and the LSD disappears.⁴

In this case, no open neural tube or LSD was identified at the time of birth. However, after the opening pressure of the lateral ventricle shunt was decreased, LSD appeared within a short period. Failure of the supratentorial ventricular system to expand after VP shunt placement, and cranial disorganization in the ectopia of the cranial gray matter may cause LSD in the membranous skull because of lack of stimulation for normal development of the calvarium. The causes of the induced LSD in this patient are unknown, but patients with similar intracranial environments have not shown development of LSD.

This case of acquired LSD indicates that abnormal disorganized collection of collagen fibers of the skull plates may be triggered by iatrogenic intracranial hypotension even after infancy.
References


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