Growing skull fracture and leptomeningeal cyst – a rare condition

A six-month-old girl was referred with a slowly enlarging pulsatile lump overlying the right half of coronal suture. She was born by ventouse-assisted delivery and the lump appeared close to the site of ventouse application. There was palpable bony defect at the base of the lump. CT brain scan with 3D reconstruction confirmed growing skull fracture and underlying leptomeningeal cyst. She underwent surgical repair of the skull defect with excellent outcome.

Linear skull fractures in young children occasionally result in a growing skull fracture (GSF). The dura mater is more adherent to the inner table of skull and therefore is easily lacerated along the fracture line. Herniation of pia and arachnoid layers (leptomeninges) occurs through the dural tear. Cerebrospinal fluid pulsations lead to progressive erosion of the skull around the fracture site. Encephalomalacia of the underlying area of brain ensues resulting in porencephalic cyst and dilatation of the ipsilateral lateral ventricle (leptomeningeal cyst).

Case study
This infant was born at our hospital by ventouse delivery as the first of a twin pregnancy. There were no antenatal concerns and no perinatal complications. Her Apgar scores were 8 and 9 at 1 minute and 5 minutes respectively. On day one swelling over the right half of the coronal suture was noted. Parents were reassured and she was discharged home shortly after. The swelling persisted and her parents noticed that it was slowly getting larger over several months. She was seen by her general practitioner who referred her for paediatric opinion.

She was seen by a paediatrician at six months of age. Her growth and development were excellent for age. She appeared very well with a small towering lump overlying the coronal suture midway between the anterior fontanelle and the right lateral end of the coronal suture (FIGURES 1 AND 2). The lump was pulsatile with a palpable bony defect at the base of the lesion (FIGURE 3). Anterior fontanelle was patent and felt of normal pressure.

Head circumference was 45 cm and showed normal growth velocity. Systemic examination was normal.
Cranial ultrasound examination through the lesion identified a moderately large cyst in the right fronto-parietal area extending from inside the lesion and inferiorly sitting on the superior wall of the right lateral ventricle. CT brain scan with 3D reconstruction confirmed diagnosis of GSF and leptomeningeal cyst (FIGURES 1-4).
She underwent repair of the skull defect using calvarially split window of normal bone (FIGURE 5). There were no intraoperative or postoperative complications. Four weeks after surgery she was in excellent health; the wound had healed well and the underlying bone appeared to be fusing as expected.

Discussion
Vacuum extraction has a relatively low complication rate and is commonly used to expedite delivery in non-progressive labour. GSF has been reported as a rare complication of ventouse-assisted delivery.

In 1997 Hes and colleagues published a case of rapid evolution of a GSF after vacuum extraction in an infant with antenatal diagnosis of spina bifida and mild hydrocephalus. They discussed that GSF is more common when the vacuum cup is applied too anteriorly thereby disrupting the coronal suture.

In 2003 Zegers and co-workers wrote up a case of GSF in a three-month-old boy following an accidental fall from the stairs on to the floor. They discussed that dural laceration is the single most important factor in the pathogenesis of GSF. They concluded that linear skull fractures in infants and young children should be monitored until definite skull bone consolidation has occurred.

Keywords
growing skull fracture; leptomeningeal cyst; ventouse-assisted delivery

Key points
1. Early recognition and appropriate imaging of growing skull fracture is required to establish the pathology.
2. Treatment should include urgent neurosurgical referral for surgical correction of the defect and organisation of careful follow-up.
In 1998 King and Boothroyd published an excellent pictorial review of cranial trauma following birth in term infants. In 1976 Rothman and colleagues published a review of the spectrum of GSF in children.

If GSF is suspected, appropriate imaging (skull X-ray, ultrasound, CT and MRI) to confirm the condition and early neurosurgical referral are very important to prevent progression and continuing damage to the growing brain.

References


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