A GSF is an uncommon (< 2%) clinical condition that can occur in infants and young children following traumatic head injury.5,7,13,14,20,22 Some authors have used the terms leptomeningeal cyst15 and cranial erosion,9,19 which describe the pathological entity but not the mechanism of action.4,10,22 A GSF requires a significant head injury, usually in infants and children younger than 2 years of age, who have a wide skull fracture, dural tear, and underlying cerebral injury.10,20,22 These injuries are noted on initial skull radiograph and CT scan. Magnetic resonance imaging will demonstrate the herniation of cerebral cortex through the fracture site.3,8 If not repaired acutely, a GSF will develop over time.2,21 Whereas GSFs have been reported in older children and young adults, they are rare in these age groups and are usually the result of an early childhood injury that is unrecognized for many years.12,16,23,24 A 1985 review of the literature of children with GSFs revealed that 34% had seizures and 38% had neurological deficits.17 It is rare for a simple linear skull fracture to become a GSF; normally it requires a wide skull fracture and underlying brain injury. Typically, on follow-up clinical evaluation after severe head trauma, the physician notes a palpable defect. Skull radiographs reveal a nonhealing skull fracture that actually widens during the period of follow-up. It often has scalloped edges, indicating bone erosion. This is confirmed by CT evaluation.

In 1977, after surgically treating 2 patients with GSFs, 1 of whom had a worsening of his mild hemiparesis, I began the policy of exploring all linear skull fractures ≥ 5 mm. This required a 1–2 cm incision over the palpable skull fracture and dural inspection by direct visualization and palpation. It was not infrequent to encounter intact dura mater. Computed tomography scanning had just become available and it became easier to evaluate the skull fracture and the degree of brain injury.11 Early exploration is still not the standard of care. By 1987, after encountering intact dura in some cases, it suggested to me that skull fractures < 5 mm did not produce leptomeningeal cysts (GSFs). It required more extensive injuries with wide skull fractures, and, in my experience, all wide skull fractures were accompanied by dural tears and often an underlying brain injury. Early repair seemed to prevent any GSF, as we had none occur at our hospital. The addition of MR imaging to evaluate the fracture for brain herniation ended the negative exploration until the occurrence of GSFs in
the present 2 cases. At this point in time we instituted a policy of delaying exploration of these wide skull fractures after encountering significant cerebral edema, which made simple dural repair more difficult. We presented our observations to the Pediatric Section Meeting of the American Association of Neurological Surgeons (Sanford RA, Ricca G, unpublished data, 1990). We later reported on a series of children. Utilizing this policy of exploring wide skull fractures, I did not encounter a child with a GSF in which the child’s primary injury was managed at Le Bonheur Children’s Medical Center from 1985 until 2007. This report will detail the errors that led to missing the injuries in 2 recent cases.

Case Reports

Case 1

This 6-month-old white male child sustained an injury in a motor vehicle accident as an unrestrained passenger. He was reported to be unconscious at the scene. By the time he arrived at Le Bonheur Children’s Hospital emergency department he was alert, recognized his parents, and had no focal deficits. Skull radiography revealed a wide diastatic skull fracture of the left lambdoid suture of approximately 5 mm at its superior portion and 4 mm at its inferior portion (Fig. 1A). Computed tomography scanning demonstrated an underlying hemorrhagic contusion just above the transverse sinus (Fig. 1B and C). Magnetic resonance imaging revealed what was thought to be herniated cerebral cortex (Fig. 1D). In keeping with the policy of delayed exploration, the patient was discharged after 3 days, doing well, and was readmitted 2 weeks later. After informed discussion with the family, the patient underwent surgery in which the dura mater was explored and appeared to be intact. On follow-up visits at 1, 3, 5, and 12 months, it was obvious that the skull fracture was failing to heal. In fact the skull fracture had actually enlarged and there was an underlying porencephalic cyst at the site of the previous hemorrhagic contusion that communicated with the lateral ventricle.

Fig. 1. Case 1. A: Lateral skull radiograph demonstrating a wide (5-mm) diastatic skull fracture. B: Axial CT scan showing a wide skull fracture at the level of diastatic fracture. C: Axial CT scan demonstrating large hemorrhagic contusion. D: Sagittal T1-weighted MR image revealing a herniated cortex and contusion.
During these clinical visits, it was noted that he had a palpable defect inferior to the site of initial surgical exploration, and his mother noted that there was a bulge overlying the skull. He was returned to surgery 7 months after the initial injury, and a craniotomy was performed, including the bony prominence that covered the site of the porencephalic cyst and the transverse sinus as a separate piece (Fig. 2C and D). There were small holes within the bone, the dura was widely separated and the porencephalic cyst extended to the bony cortex. A dural patch graft of bovine pericardium was sutured in place and bone replaced (Fig. 2E and F).

Postoperatively, the child has done well. It was apparent on close review of this case that the initial surgical exploration via a linear incision was of the widest portion of the skull fracture superiorly where the dura was intact, and the inferior portion, where the dura was torn, was neglected. The MR imaging abnormality was at the site of the contusion and probably was real in retrospect but was dismissed after initial exploration revealed intact dura.

Case 2

This 14-month-old white male child fell approximately 40 ft and landed on a concrete surface. He was unconscious at the scene. On arrival to the emergency department, he was moving all extremities, his eyes were open, he did not attend to his surroundings, but there was no focal neurological deficit. Skull radiography (Fig. 3A) revealed a wide skull fracture that extended posteriorly from the coronal suture, was levered up posteriorly (Fig. 3B), and extended to the midline. Computed tomography scanning revealed an underlying cerebral contusion and a small acute subdural hematoma (Fig. 3C and D). The patient was admitted to the intensive care unit and his case was managed expectantly. He had significant pulmonary issues and remained intubated for 1 week, his condition was stabilized, and the patient was moved to the floor.

Magnetic resonance imaging (Fig. 3E and F) demonstrated a herniated cerebral cortex. The child was discharged from the hospital 12 days postinjury, readmitted, and taken to the operating room 17 days after injury. A linear skin incision at the posterior portion (widest point) of the fracture was made. Inspection of the dura revealed it to be intact.

On follow-up clinic visits at 4 and 6 months, it was obvious that the fracture was not healing and had actually enlarged. Computed tomography scanning revealed peculiar eburnated bone and a wide skull fracture (Fig. 3G and H). Because of the GSF, surgical repair was undertaken. Bur holes were placed laterally and 2 bone flaps were removed on either side of the wide fracture (see the following section). The dura was widely retracted with the herniated cerebral cortex. The dural margin was dissected toward the midline to the sagittal sinus. As the dura was gently teased away from the bone, significant bleeding was encountered. Further bone was removed to completely expose the sinus. Bleeding was coming from a dorsal rent approximately 1.5 cm in length. The dura was repaired by making a patch of gel foam covered with Surgicel and suturing it across the sinus. The impression at surgery was that this dural defect...
was due to the original injury and had healed by fusion to the bone, not by true dural repair. The child made an uneventful recovery.

On careful review of this case, we found it obvious that the initial failure to appreciate the dural tear was an error produced by exploring the most widened area of the fracture, which was posteriorly located, and ignoring the hemorrhagic contusion anteriorly where the bone had levered inward, tearing the dura and lacerating the cerebral cortex (Fig. 3C and D) and not correctly correlating the injury on MR imaging with the skull radiography.

Technical Note: Acute or Delayed Repair of Wide GSFs

The scalp incision needs to be carefully planned to allow for adequate bony removal and dural repair should a dural defect be encountered. The first step in the acute operation is to confirm the presence of herniated cerebral tissue, because if no dural defect is encountered the procedure can be terminated at that point. Once cerebral tissue is noted, one can anticipate that dura has retracted 1–2 cm beneath the intact bone. At this point bur holes are made on either side of the skull defect (Fig. 4 upper). When dura is encountered, the bone flap is mobilized to the skull fracture; if not, a second hole is made further away until intact dura is noted before removing the bone. This maneuver is duplicated on the opposite side of the fracture. The surgeon now has 2 bone flaps to be replaced after dural repair (Fig. 4 lower).

The edge of the dura is carefully freed from the herniated cerebral tissue with sharp dissection and bipolar coagulation. Mannitol and/or Lasix are useful to facilitate brain relaxation. If the surgery is performed during a relatively short time after injury, then the cortex will not be attached to the scalp. In the cases in which surgery is performed after GSF development, the herniated cortex will have parasitized blood vessels from the scalp, and dissection is very difficult. Viable cortex may be injured even with the judicious use of bipolar coagulation because the gliotic brain is so adherent, similar to the difficulty encountered with reoperation for recurrent tumor. This may be avoided by acute repair because there is no need to use diathermy to separate brain from torn dura.

It is extremely important that the approximate location of the venous sinus be drawn on the scalp so that bone flaps can be placed either to avoid the sinus or to adequately expose it in anticipation of necessary surgical repair—always a challenge in a young child. As dural dissection proceeds toward a venous sinus, adequate bony exposure is critical. Most dural tears extend to the sinus, which limits further extension, but one must be prepared to treat the true rent in the sinus. The technique of drilling or biting bone to the margin of the dural sinus in a piece-meal fashion and tacking the edge to the bone works, but it is less controlled if bleeding is encountered and complicates the process of approximating the bone over the dural repair. If there is a tear in the dura, vigorous bleeding is encountered and must be controlled quickly. I have found that a piece of Gelfoam wrapped in Surgicel makes an excellent patch. It is held in place with gentle pressure. At this point, primary suture of the rent may be attempted, sliding the patch as a running locking stitch is used. If the dural margin is adequate, this can be accomplished with minimal blood loss. If the dural edges do not lend themselves readily to repair, place sutures in good dura on either side of the patch and tie them over the Surgicel-wrapped Gelfoam, securing it in place. Once the bone is replaced and dural tack-up sutures are placed, adequate closure has been achieved.

After the dural margin is developed, the dura may be repaired with a patch of the surgeon’s choice. This may be temporalis fascia, lyophilized cadaver dura, or bovine pericardium (our choice). I do not recommend synthetic materials, and the use of a fascia lata graft from the thigh is no longer necessary since the substitutes work so well.

After a watertight closure is accomplished, dural tack-up sutures are placed to secure the patch to the bony margin. The 2 bone flaps are approximated in the midline.
Prevention of growing skull fractures

over the dural patch with any residual bony defect laterally over normal dura (Fig. 4 lower). In the young child and infant, new lateral bone growth results in no residual skull defect. It is important that bone fixation be rigid, as pulsation will result in excessive resorption of the bone, requiring cranioplasty at a later date.

Discussion

The pathogenesis of the GSF is thought to be a combination of factors in the majority of cases.20 The first is a significant head injury and wide skull fracture in which the dura mater is torn and cerebral tissue herniates through the dural defect. This injury is readily identified by MR imaging confirming the seriousness of the injury.2,3,8 Experimental work in puppies by Goldstein et al.6 has demonstrated that for a GSF to occur the arachnoid has to be torn; a dural tear alone was insufficient to produce bone erosion. The addition of brain injury did not increase the incidence of GSF.

We coined the term “burst skull fracture,” which we believed described the most severe of these injuries.2 In younger children with thinner bone, significant traumatic force results in dural and bone disruption. A secondary factor is the growing child’s brain, which pulsates through the dural and bony defects and erodes the bone, increasing the width of the fracture and creating the characteristic scalloped edges.5,13,19,22 Vascularity from the brain is parasitized by the scalp, which is now in direct contact with the cerebral cortex, and this may increase the chance of delayed seizures.

Exploration of wide linear skull fractures following acute head injury in infants and repair of the dural tear is worthwhile to prevent GSFs.2,21 This is suggested by the experience of this author’s failure to encounter a GSF in 35 years of care in busy trauma services at 2 children’s hospitals. This series was broken by technical errors in surgery when possible allows the cerebral edema to resolve and permits surgical intervention in a hemodynamically stable child. Venous sinus injuries can be better predicted and repaired in a controlled fashion. Most importantly, whenever possible. Acute epidural, subdural, and/or intracerebral hematoma may require acute intervention and only rarely is a decompressive craniotomy necessary to control severe brain edema. A delay in surgery when possible allows the cerebral edema to resolve and permits surgical intervention in a hemodynamically stable child. Venous sinus injuries can be better predicted and repaired in a controlled fashion. Most importantly, this early dural repair and replacement of bone will prevent the GSF. Numerous authors have described the late onset of seizures, progressive neurological deficit, ventricular porencephaly, and leptomeningeal cyst occurring in a delayed fashion as the GSF occurred.4,5,7,13,14,18–20,22 All of these late effects of GSF can be prevented by early surgery. Especially disheartening is the child who has a minor neurological deficit prior to GSF surgical repair who develops significant dysfunction following repair from injury to functional cortex.

Exploration of wide linear skull fractures needs to proceed after careful interpretation of the imaging. In fact, skull radiographs may be easier to interpret than CT scans in terms of helping to locate the area of dural tear. When an MR image demonstrates definite leptomeningeal tissue, it needs to be carefully correlated with the skull fracture noted on the CT scan and skull radiograph. The site of brain herniation is the area that needs to be explored. The 2 cases described in this report illustrate the error of finding intact dura due to poor interpretation of the neuroimages.

Conclusions

Head injuries in infants and young children accompanied by wide skull fracture predict dural laceration. These injuries are usually associated with some degree of cerebral injury (contusion and/or hemorrhage). Magnetic resonance imaging demonstrates the herniation of cerebral tissue through the dural defect. Repair of the dural tear acutely prevents the development of GSF and secondary cortical injury (that is, additional neurological deficit and seizures). This repair is best delayed until the child is hemodynamically stable and cerebral edema has resolved.

Disclosure

The author reports no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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