GROWING SKULL FRACTURES OF CHILDHOOD*

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A fracture of the skull occurring in infancy or childhood ordinarily heals without difficulty. Rarely, however, such a fracture enlarges progressively to form a permanent cranial defect. Common features that constitute this disorder serve to identify it as a unique entity.

Important clinical and pathological observations, particularly those reported before the turn of the century, have not been appreciated in recent case reports dwelling on various aspects of this syndrome. Consequently, we have made a comprehensive survey of world literature in an effort to clarify the clinical picture and gather together common features that bear on pathogenesis. In addition, personal observations on a series of 5 cases are presented.

HISTORICAL REVIEW

The first report of this condition was in 1816 by John Howship who noted “partial absorption of the (right) parietal bone, arising from a blow on the head” in a child aged 9 months. Leeches were applied to the temples directly after the head injury. Within a fortnight a pulsatile defect was perceived at the site of injury and shortly afterward a left hemiparesis was apparent. When seen at age 4, “the left side was still deficient . . . while the opening in the cranium remained undiminished. Upon laying the hand on the part, the pulsations of the brain were felt strong and distinct.” Howship ascribed the lesion to a local debility in cranial circulation.

Of prehistoric note is the suggestion of Alajouanine and Thurol that certain ancient skull openings, presently interpreted as primitive trephinings, actually may be defects that followed fractures of infancy.

The first account of the pathology was made in Vienna in 1856 by Rokitansky, who described the autopsy findings in a child, aged 8 months, with a cranial aperture which measured 6 x 4 cm. Above this defect was a fluid-filled sac, and beneath it was a dural opening and an underlying brain injury. The clinical features, later elaborated by Weinlechner, were that the child was normal until the age of 5 months when a fall associated with a convulsion was followed by an enlarging cephalhydrocele. Puncture of the sac was followed by symptoms of meningitis and subsequent death.

In 1862 Billroth made a detailed case report in which the term “meningocele spuria” was used to describe the external sac of cerebrospinal fluid. Injury occurred from a forceps delivery, and a few days after birth a bulging appeared beneath the scalp on the right side. This gradually grew in size until eventually it became nearly as large as the head itself. Billroth saw the child at the age of 2 ½ years and advised puncture of the sac. This procedure emptied it of a clear fluid and thereby revealed a palpable underlying bone deficiency. Death followed injection of iodine into the sac. At autopsy an opening in the dura mater was seen to correspond to the cranial defect and there was a direct communication between the “meningocele spuria” and the lateral ventricle.

In most of the earlier reports the principal
feature of this condition was considered to be a collection of cerebrospinal fluid external to the skull. If subsequent loss of cranium at the site of fracture was noted, it was given only secondary attention. Thus the disorder was described, particularly by British authors, as a fracture of the skull followed by a collection of cerebrospinal fluid beneath the scalp. The title traumatic cephalhydrocele was used in 1884 in the first American report which was by P. S. Conner of the Medical College of Ohio. Sir Thomas Smith used the same term, but both authors presented cases in which the late appearance was that of a pulsatile cranial defect and not a cephalhydrocele.

Godlee, in 1885, reported 2 cases of skull fractures followed by pulsating subcutaneous tumors. Although his title referred to the cephalhydrocele, his grasp of the total clinical picture is apparent in his superb introduction:

“The fracture referred to can apparently, only occur in young children; it is a simple fracture and is accompanied at the time of the accident by what appears to be an ordinary haematoma beneath the periosteum; but this haematoma is noticed either immediately or after the lapse of a certain time to pulsate. Eventually it is found that there is an aperture of considerable size in the skull, and, if the child dies, not only is the dura mater discovered to be ruptured, but in many, if not in all cases, the brain itself is seen to have suffered a serious injury.”

Concern with the cephalhydrocele began to wane and consideration of the skull defect as the significant aspect of this syndrome appeared in the German literature with the report of Weinlechner in 1882. This was followed in 1885 by a report by von Winiwarter of a large hiatus in the skull which followed head injury in infancy. Henoch, following this trend, reported 2 cases in 1888 under the title “Über Schädel-lücken im frühen Kindesalter.”

Neurosurgical consideration was given in 1907 by Sir Charles Ballance in a report of an operation on a patient, aged 4, who had a head injury, subsequent cephalhydrocele, widened fracture, dural defect and underlying brain injury. Sir Wilfred Trotter operated on a patient in whom a local cystic dilation of the lateral ventricle extended out to the cranial defect. On the basis of this observation he chose to entitle the disorder “traumatic ventricular cyst,” even though he noted three characteristic features of such cases to be gap in the skull, pulsation, and history of birth or childhood injury.

About the turn of the century, when the use of roentgen ray appeared as an aid to diagnosis, there was, oddly, a decrease in reports of this condition. The roentgenographic aspects of this disorder were outlined by Dyke in 1937 without reference to previous pathological or clinical observations. He used the title “leptomeningeal cysts,” which is a significant term because of the profound influence it has had on subsequent case reports. It was stated that, “Leptomeningeal cysts which result from trauma occur with equal frequency in children and adults. The trauma, as a rule, is severe and is accompanied by evidence of skull fracture.” It was theorized that, “loculated, fluid-filled spaces form cysts and, owing to the pulsations of the brain, the overlying bone is absorbed.” The roentgenological diagnosis was based on (1) widening of an old fracture, (2) scalloping of the bordering inner table of the skull, and (3) localized increase in vascularity of the bone. An illustration showed such a widening of a fracture in a child’s skull, and that case has since become celebrated by appearance in no less than six different publications.

Dyke’s statement that these lesions occur with equal frequency in children and adults suggests that he was considering more than one condition, since enlargement of fractures occurs only in children. The embodiment of a hypothetical concept of pathogenesis in the term “leptomeningeal cyst” has been a confusing influence on subsequent case reports.

Pancoast et al. in 1940 used the term “fibrosing osteitis” which was based on pathological study of biopsy material. They considered the cause to be disturbance in blood supply secondary to injury. In their later work, Pendergrass et al. continued to