Growing fractures of the skull

DEREK KINGSLEY, KENNETH TILL, AND RICHARD HOARE

From The Hospital for Sick Children, Great Ormond Street, London

SUMMARY Ten cases of growing fractures seen in the last 10 years are presented. Six of the patients sustained their injury within the first six months of life. The defects formed rapidly, several within two or three months after injury. There was enlargement of the defect in only one case after the date of discovery. Although the defects involved the parietal bone most commonly, in four out of 10 the lesion crossed either the coronal or the lambdoid suture. The edges of the defects were usually thickened; in some areas they were saucer-shaped but in two cases there was erosion of the outer table of the skull at a distance from the margin of the defect, the erosion being related to an extracranial fluid-filled cavity in continuity with a porencephalic cyst. The ipsilateral ventricle was usually dilated and in a number of cases was associated with a porencephalic cyst. In no case was a "leptomeningeal cyst" found beneath the defect at operation. A detailed review confirms many of the findings previously described but suggests that the pathology of the condition is still not fully understood. Computed tomography, undertaken in one case, appears to be the examination of choice. Further light may be thrown on the pathogenesis of this condition by the use of intracystic, intrathecal, and intraventricular water-soluble contrast media.

Growing fractures of the skull are rare and almost unknown after the first few years of life. The term "growing fracture" was first used by Pia and Tönnis (1953) in a major study published in the German literature. Before this publication, the condition went under a number of different names of which leptomeningeal cyst was probably the best known (Dyke, 1938). In the same year Taveras and Ransohoff (1953), in a study of seven cases treated by surgery, suggested that the single most important factor in its pathogenesis was a dural tear. This, together with arachnoid adhesions aided by normal pulsations of brain and perhaps a ball valve mechanism, produced the enlarging bony defect.

In 1961 Lende and Erickson first reviewed the world literature and reported clinical details of three cases. They emphasised that there were four essential features: (1) a skull fracture in infancy or early childhood, (2) a dural tear at the time of fracture, (3) brain injury beneath the fracture, and (4) subsequent enlargement of the fracture to form a cranial defect.

However, while these features form an essential part of the "syndrome", they are simply a mani-

festation of the severe degree of trauma to the skull and brain occurring at a time of maximum growth of both cranial vault and underlying brain.

At this hospital we have encountered only 10 cases in the past 10 years. These cases have been extensively investigated and have enabled us to confirm most of the previously documented features, but our observations suggest that leptomeningeal cysts probably do not exist in this condition or, if they do occur, are very rare. Furthermore, the changes in intracranial pathophysiology together with the dural tear are probably sufficient for the development of such a defect. Case reports of only two children are presented, both of whom had unusual features.

Case reports

CASE 1

In February 1975, at the age of 3 years and 5 months, the patient fell 5 m from a school window. On admission to hospital she had bilateral periorbital haematomas, and skull radiographs revealed a left frontoparietal and left temporal fracture.

Six months after this episode she was noted to have a collection of fluid under the scalp in the right frontal region associated with a palpable

Address for reprint requests: Dr D. Kingsley, Lysholm Radiological Department, National Hospital, Queen Square, London WC1N 3BG, England.

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bony defect. The soft tissue swelling had refilled immediately after aspiration at another hospital.

Skull radiographs showed a left frontal defect measuring 80×15 mm with extensive thinning of the outer table extending across the midline and involving much of the right frontal bone (Fig. 1). A pneumoencephalogram showed a bilocular porencephalic cyst connecting with a dilated frontal horn of the left lateral ventricle. The third ventricle was displaced to the same side. No air was seen over either hemisphere.

At operation, atrophic brain was seen underlying the defect. The dura mater was absent and the periosteum thickened but adherent to underlying arachnoid mater. To the right of the defect there was erosion of the outer table of the frontal bone with a fluid filled space lying deep to the pericranium and continuous with the intracranial cavity (Fig. 2). A rib graft was used to close the defect and the patient made a satisfactory recovery. Two and a half years later she remains well with no neurological deficit.

CASE 2

At the age of 3 months the patient was involved in a road traffic accident. The child had a grossly comminuted fracture of the left parietal region extending across the midline with a severe right hemiparesis which, however, showed subsequent improvement. At 5 months of age a pulsating defect was noted in the right parietal region, but the parents reported that the defect had been enlarging from the time of injury. The child was found to have a squint. Skull radiographs showed a large defect in the left parietal region with eleva-

Fig. 2 Operative diagram (case 1) demonstrating the connection between the subpericranial fluid collection, and porencephalic cyst, and the dilated left lateral ventricle.

tion of the neighbouring skull vault. A pneumoencephalogram showed a dilated left lateral ventricle, particularly the trigone. The ventricles were displaced to the left, and it was thought that a cyst was probably present under the defect. Air did not reach the sulci on either side.

At operation, cerebral cortex was found in the defect and was firmly adherent to the overlying periosteum. An underlying porencephalic cyst was present but did not communicate with the ventricle, and an opening was, therefore, made between the two to promote drainage. Post-operatively, a subcutaneous collection of cerebrospinal fluid developed over the defect, and subsequent films suggested the development of local erosion of the outer table of bone immediately beneath the extracranial cyst (Fig. 3).

Fig. 1 (a) Posteroanterior skull radiograph of case 1 showing left frontal bone defect with thinning of the outer table extending across the midline to the right side. (b) Tangential view of the defect showing erosion of the outer table.

Fig. 3 (a) Postoperative appearances (case 2) demonstrating air in a cavity outside the vault with overlying soft tissue swelling. (b) One month later. The extracranial cyst remains, and there is now erosion of the outer table immediately beneath. There is widening of the sutures due to raised intracranial pressure. (c) Three months later. The soft tissue swelling is smaller, the sutures are now normal, and the erosion of the outer table has become smooth.

Pneumoencephalography showed further dilatation of the left lateral ventricle which connected through the defect with the extracranial cavity. A ventriculoatrial shunt was inserted but required numerous revisions over the next eight years. However, for the past two years her condition has remained stable. She attends a special school and continues to develop satisfactorily.

Analysis of cases

A summary of the clinical features is shown in the Table.

AGE OF CHILD AT TIME OF INJURY

Two cases sustained birth trauma and the oldest patient in this series was aged 3 years and 5 months. Six of the 10 patients sustained their injury within the first six months of life, and all but one within the first 18 months of life.

DEVELOPMENT OF BONE DEFECT

The shortest period between the head injury and the discovery of the cranial defect was about two months. This occurred in the patient who was savaged by a dog. In one other patient the defect had developed within three months of injury, although in both of these patients the defect appeared to have enlarged gradually throughout this period. In two other patients the soft tissue swelling appeared three weeks after the injury.

NATURE OF DEFECT

Most of the bone defects were of considerable size, the largest one measuring 100×70 mm. In one case there were bilateral defects. Six of the defects

were confined to the parietal and one to the frontal bone. Two were frontoparietal and one parieto-occipital but did not reach the posterior fossa. Defects crossed the sutures in four of the 10 cases. The defects were normally irregular in shape and, in some cases, parts of the edge were elevated. There was marked thickening of the edges particularly when the defect was of long-standing.

INTRACRANIAL ABNORMALITY

Eight of the 10 patients had pneumoencephalography. In each case there was dilatation of the ipsilateral ventricle and in five of these there was also displacement of the third ventricle to the affected side. In seven patients there was an associated cyst. Six of these cysts were porencephalic and four communicated with the lateral ventricle. Leptomeningeal cysts were not demonstrated in any of the seven cases explored.

In two cases air was present in the sulci of the uninvolved hemisphere, but did not reach the convexity of the affected side (Fig. 4). In no case did air in the subarachnoid space fill any cavity in the region of the defect, a finding which was explained at surgery, where the meninges and underlying brain were seen to be adherent to each other in and around the bone defect.

One patient was investigated by computed tomography (CT) alone, and surgery was undertaken without any invasive diagnostic procedure. Computed tomography demonstrated the bone defect as well as the underlying ventricular dilatation and cystic cavity (Fig. 5). It is impossible to assess the presence or absence of continuity with the ventricle without the use of intraventricular or

Table Summary of clinical details in 10 patients with growing fractures of the skull

Case number	Age at time of injury (mo)	Cause	Age at diagnosis of defect (mo)	Symptoms	Site	Size (mm)	Suture involvement	Position of third ventricle	Dilation of lateral ventricle	Porencephalic cyst	Hemisphere involved	Bone defect explored
1	41	Fell 5m	23	Swelling	L. frontal	80 x 15	—	To the left	+	+	—	—
2	3	Motor accident	5	R. hemiplegia	L. parietal	70 x 50	—	To the left	+	+	—	+
3	6	Fell 3m	23	Swelling Squint	R. parietal	50 x 20	—	Midline	+	—	—	—
4	1	Nonaccidental injury	23	R. hemiplegia	L. parietal	70 x 40	+	To the left	+	—	—	—
5	16	Savaged by dog	17	Swelling	Biparietal	50 x 50 30 x 60	+	Midline	+	+	+	+
6	1	Fell 3m	23	Swelling	L. parietal	75 x 15	—	Midline	+	+	+	+
7	1	Fell 1m	18	Convulsions	L. parietal	100 x 70	—	To the left	+	+	—	+
8	Birth	Forceps injury	3	Swelling	L. fronto-parietal	40 x 50	+	To the left	+	+	+	+
9	Birth	Forceps injury	4	None	L. fronto-parietal	45 x 45	+	Midline	+	—	—	—
10	9	Nonaccidental injury	72	R. hemiplegia	L. parietal	100 x 50	—	Midline	+	+	—	+

Fig. 4 Pneumoencephalogram demonstrating many typical features: dilatation of the ipsilateral ventricle towards the defect; displacement of the third ventricle to the left; air in cortical sulci on the right but not on the left; thickened sclerotic margin of the defect with irregular bone erosion and expansion.

Fig. 5 CT scan (case 10). Serial slices demonstrating the bone defect, dilatation of the left lateral ventricle with overlying low attenuation probably due to a cyst, and some left hemisphere enlargement.

intracystic contrast media, and such studies may throw further light on the pathogenesis of this condition. At present, however, CT gives sufficient information for the management of the patient and, where available, is the examination of choice.

Discussion

Most of the postnatal growth and differentiation of the brain and skull vault occurs within the first two years of life, and by the end of this time the inner and outer tables, diploic spaces, diploic veins, vascular markings, and grooves on the internal surface of the calvaria are all present (Caffey, 1967).

In their review of the world literature up to

1960, Lende and Erickson (1961) reported that over half of the fractures occurred under the age of 12 months and that 90% of them occurred under the age of 3 years. Our series agrees with these findings, two-thirds of them occurring before the age of 1 year, and all but one under the age of 3 years.

The dura mater consists of two layers, the outer one being in close contact with the bone and periosteum. In childhood this relationship is even closer (Schwartz, 1941) which may account for the rarity of extradural haematomas at this age. It is not surprising, therefore, that major diastatic fractures of the bones of the vault are likely to cause tearing of the dura mater.

A notable feature of this condition was the lack of neurological symptoms in our patients. Despite the severe nature of the injury and the very considerable amount of underlying brain damage demonstrated by pneumoencephalography, CT scanning, and at operation, only one patient had generalised convulsions, three had mild hemiplegia, and one had a left convergent squint. Follow-up has continued on eight of the children up to the present, the total period being up to 10 years. Two children still have a mild hemiparesis and one is slightly backward but the remainder are normal.

After the initial injury these fractures enlarge rapidly, frequently reaching their full size within two months. Further absolute enlargement of the bone defect may occur over a longer period, and has been encountered recently in one untreated patient (case 9, Table) radiographed again two years after initial diagnosis. The intracranial state has not been documented up to the present.

Although a dural tear with severe underlying brain damage is necessary for the formation of a growing fracture, uncertainty remains as to why the defect should enlarge and, having reached a certain size, why it should cease to grow. In 1967 Goldstein *et al.* published an experimental study on enlarging skull fractures in young dogs. They found a high incidence of enlarging bone lesions in those groups where the defect was not limited to the dura mater alone. However, in the group with a dural lesion but without damage to the arachnoid or pia mater, or to the underlying brain there was no significant incidence of growing fracture. Similar experiments by Lende (1974) suggested, however, that bone defects do not heal where there is an underlying dural defect, and that underlying brain damage does not alter the bone defects. However, simulation of the natural pathological state is extremely difficult to achieve and some doubt must remain as to the value of these experiments since the degree of intracranial damage *in vivo* on both the ipsilateral and contralateral sides is likely to be more severe than that existing in the experimental state.

Hakim (1973) has shown that any increase in CSF pressure due to partial blockage of the drainage pathway or overproduction of CSF will tend immediately to compress the veins carrying blood out of the cranial cavity, and will lead to ventricular enlargement. When the ventricles are small, quite large changes in CSF pressure are possible with little change in the stress at the subdural level due to the elastic properties of the brain tissue which reduce the stresses transmitted through it. However, when ventricles are large any increase in CSF pressure brings about an im-

mediate and almost equal increase in subdural stress. It can be assumed that, after an injury to the cranial vault of such magnitude as is required to produce a growing fracture, the normal equilibrium of production and absorption of the cerebrospinal fluid is temporarily upset. Fairly rapidly, however, a new equilibrium is set up dependent upon changes in the ventricular size, the development of a porencephalic cyst, and the healing processes of the damaged layers, at which stage no further change is likely to occur in the size of the defect.

The shape of the defect depends on the relationship between the forces tending to enlarge the defect and those tending to produce healing of the skull fracture. Most defects are irregular and scalloped. In some patients areas of rarefaction and expansion coexist with markedly thickened edges. Occasionally a saucer-like appearance is seen with greater erosion of the outer table of bone; but it must be rare for a large defect to occur in the outer table as happened in case 1 where persistence of a defect in the pseudodura resulted in the development of a subgaleal cavity in continuity with the intracranial porencephalic cyst. The intracranial pressure was, therefore, transmitted into the subgaleal fluid collection with subsequent pressure erosion of the outer table. That such erosion of the outer table can occur at a distance from the defect is further reinforced by case 2. After surgical exploration of the defect, a cystic space developed subpericranially in continuity with the porencephalic cyst. Postoperatively, the patient's course was stormy with evidence of raised intracranial pressure, and follow-up films suggested local erosions of the outer table beneath the cyst. It is likely, therefore, that transmission of pressure through a cystic cavity can produce erosion of the outer table of the skull adjacent to the fracture site in much the same way as does a pulsatile aortic aneurysm of the dorsal vertebral bodies.

Dyke (1938) was the first to use the title "leptomeningeal cyst" when he suggested that after severe trauma with skull fractures "loculated fluid filled cysts develop and owing to pulsations of the brain the overlying bone is absorbed."

Later Taveras and Ransohoff (1953), in their study of seven cases, suggested that such cysts formed between pia and arachnoid membranes, and that there was herniation of arachnoid mater through the defect. In all their cases they found cystic structures beneath the defects but the exact position of these cysts is uncertain. No comment was made on the relationship of the cysts to the surrounding brain tissue. Lende and Erickson

(1961) referred to the term "leptomeningeal cyst" but noted that they had not encountered any such cyst-like structures in their cases. They suggest, however, that such cysts may sometimes occur after local injury, but, in a later paper, Lende (1974) considered such cysts to be rare while Penfield, in a discussion of a paper by Caffey (1954), suggested that there might be brain rather than a leptomeningeal cyst between the edges of the skull defect. Our experience confirms that of Lende and Erickson and of Penfield. At operation the pia and arachnoid membranes were matted together and stuck both to the edge of the defect and to the underlying atrophic brain. The edges of the defect were abnormally thick and the intervening area usually filled with fibrous tissue. In no case was there any evidence of a leptomeningeal cyst. Occasionally cysts have been described situated between the fibrous tissue and underlying cortex, but these are small and rare, and it seems inconceivable that they can contribute to the enlargement of the defect. It appears, therefore, that "leptomeningeal cysts" are very rare and probably of little importance, and that almost all the cysts that are demonstrated are porencephalic, related to the severe brain trauma, whether or not they communicate with the ventricles.

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