CASE SERIES

Growing skull fractures in a developing country: a case series

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ABSTRACT

Background: Growing Skull Fracture (GSF) is a very rare complication of skull fractures. Different types have been described and because presentation may be delayed with neurological sequelae, diagnosis and prompt intervention are key to achieving good outcome.
Objective: This study seeks to highlight the features and the technical peculiarities in management of growing skull fractures in low resource settings.
Methodology: A retrospective study of patients who were managed in Ahmadu Bello University Teaching Hospital, Zaria, Kaduna State, Nigeria, January 2006–October 2015. Relevant data were retrieved from patients' medical records and analyzed.
Results: Seven patients were managed over the period under review, but complete records were found in only 4 cases. The age range was 6weeks-48years, and 6 patients (85.7%) were in the paediatric age group (6weeks-16months), with a mean age of 6.25months. All the paediatric patients were less than 2years of age. Three patients (42.9%) were females and four (57.1%) were males, giving a M: F ratio of 1.3:1. The aetiological factor in six of the patients (85.7%, all the paediatric cases) was road traffic accident. One patient (14.3%, the only adult) was assaulted, had a porencephalic cyst (type III GSF). One other patient had type-I GSF and was treated with cysto-peritoneal shunting, five had type-II GSF, four were managed operatively with craniotomy and duroplasty, while, one was lost to follow-up. Surgical outcomes were adjudged as good in all patients, but, anaemia was a problem in the paediatric patients.
Conclusion: Growing skull fracture is predominantly a disease of the paediatric age group which may, nevertheless, be seen in adults. Prompt and appropriate surgical intervention has a good outcome.

***Keywords*:** Craniotomy, duroplasty, good outcome, surgical management

INTRODUCTION

Growing skull fracture (GSF) is a very rare complication of linear skull fractures in young children (usually younger than 3years).[1] Lack of consensus on the cause is reflected by the different names used to describe it: traumatic ventricular cysts, cranio-cerebral erosions, cranial malacia, post-traumatic leptomeningeal cysts, traumatic meningocele, cephalhydrocele, spuria, among others. It consists of a fractured line that widens with time and usually requires both a widely separated fracture and dural tear to occur. Though predominantly seen in children, cases have been reported among adults.[2,3] Left unattended to, secondary brain injury have

been observed months to years after the initial injury due to local brain herniation and consequent ischaemia, repetitive trauma to the exposed brain on the bone edge, or by direct concussion and physical distortion of the brain related to its displacement.[4] Three types of GSF have been described: Type I - Leptomeningeal cyst, Type II - Damaged and gliotic brain, and Type III - Porencephalic cyst.[5] Identification and appropriate treatment are, therefore, imperative in order to limit neurological sequelae.

METHODOLOGY

A retrospective study of patients who were managed for Growing Skull Fractures in Ahmadu Bello University Teaching Hospital, Zaria, Kaduna State, Nigeria, January 2006–October 2015 was conducted. Patients' medical records were retrieved and age, gender, clinical presentation, radiological and laboratory findings, treatment and outcome were documented and analyzed.

RESULTS

Seven patients were managed over the period under review. Complete records of 4 of the 7 patients were found. The age range was 6weeks-48years. Six patients (85.7%) were in the paediatric age group (6weeks-16months) with a mean age of 6.25months, and all the paediatric patients were less than 2years of age. Three patients (42.9%) were females and four (57.1%) were males giving a M:F ratio of 1.3:1. The aetiological factor in all the paediatric cases 6 (85.7%,) was road traffic accident. One patient (14.3%, the only adult) was assaulted; *see Table 1.0.*

Table 1.0. Summary of patients with growing skull fracture

Case	Sex	Age	Site	Hb*(g/dl)	Type of GSF	Aetiology
One	Female	6 weeks	Parietal	5.7	Type II	RTA**
Two	Male	8 weeks	Parietal	6.0	Type I	RTA
Three	Male	8 months	Fronto-Parietal	6.3	Type II	RTA
Four	Male	48 years	Occipital		Type III	Assault
Five	Female	16 months	Temporal	-	Type II	RTA
Six	Male	7 months	Parietal	-	TypeII	RTA
Seven	Female	3 months	Parieto-Occipital	-	-	RTA

**Haemogram* ***Road Traffic Accident*

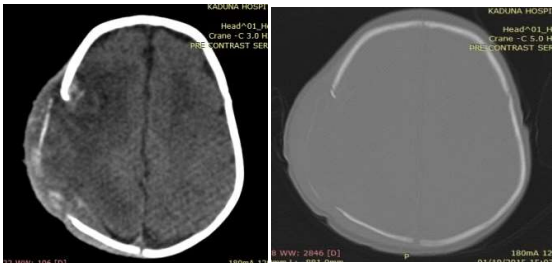
The 6 paediatric patients presented between 4hours and 4weeks after their incidents, while, the only adult patient presented 14years after. Scalp swelling was noticed 90minutes–16hours of the incident and was present in all 6 paediatric patients.

Case 1

A 6-week old girl was on the back of her mother on a motorcycle when they were knocked down by another motorcycle. The patient was noticed to have a right scalp swelling on the same day of the incident and was treated on out-patient basis in a peripheral hospital. One week later, she was referred to our centre on account of seizures and progressive increase in the size of the scalp swelling. Significant examination findings included pallor, a soft pulsatile scalp swelling on the right fronto-parietal region measuring 14x10cm, overlying a palpable skull defect. Haemogram was 5.7g/dl, and correction of the anaemia was achieved after transfusion of 3units of blood.

A diagnosis of a growing skull fracture was confirmed with brain computed tomography, *see Figures 1a and b.*

Figures 1a and 1b showing diastatic fracture, brain swelling and herniation



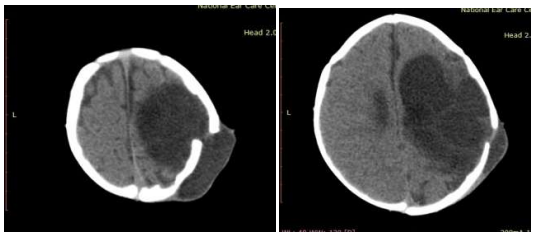
Case 2

An 8-week old male infant presented with multiple injuries following RTA while he was riding with his mother on a motorcycle. He presented with loss of consciousness, right scalp swelling and right femoral fracture within 24hours.

Examination revealed a GCS of 3, pallor, dilated unreactive right pupil (7mm), left hemiparesis and right closed femoral fracture. He also had a soft, pulsatile, left parietal scalp swelling overlying an area of skull defect and measuring 12x10cm.

Brain CT scan confirmed a GSF (Figure 2), and haemogram on admission was 6.0g/dl. He was co-managed with Orthopaedic surgeons, and had antibiotics and anticonvulsants then, subsequently, had a cysto-peritoneal shunt with complete resolution of the swelling.

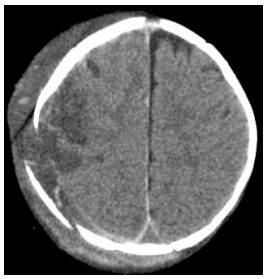
Figure 2. Left parieto-occipital scalp swelling, skull fracture and contused brain on CT



Case 3

An 8-month old male infant was admitted 16 hours after involvement in a passenger motor vehicle RTA with transient loss of consciousness and right scalp swelling. He was found to be pale, and had a soft pulsatile scalp swelling involving the right fronto-parieto-occipital region and overlying a palpable skull defect. He, also, had hypertonia and hyperreflexia in the left upper and lower limbs. Brain CT scan confirmed a GSF, *see Figure 3*. Haemogram was 6.3g/dl. He had blood transfusion to correct anaemia and had anticonvulsants and antibiotics. He eventually had craniotomy and duroplasty with good outcome.

Figure 3. Right diastatic skull fracture with herniated contused brain tissue on CT



Case 4

A 48-year old business man presented to our service with visual deterioration and diplopia of 2years' duration. He, also, complained of seeming inability to carry out learned tasks such as driving, and had agraphia. No hemibody-neglect or personality changes. No headache, convulsion or paresis. He was assaulted 15years prior to presentation and sustained machete cuts and was managed for severe head injury with elevation of depressed segment. He did well and was discharged home. He remained in good state of health until the present symptoms were noticed. He was not a known hypertensive or diabetic, and had a GCS of 15, with no obvious cranial nerve palsy.

Brain CT Scan showed a 4.2cm skull defect in the left occipital area and a hypodense non-enhancing area of CSF density in the left occipital lobe communicating with ipsilateral posterior horn of the lateral ventricle measuring 6.1x4.7cm; *see Figures 4a and b.*

A diagnosis of left occipital porencephalic cyst was made and the patient was managed non-operatively.

Figures 4a and b. Brain CT showing a left occipital skull defect with porencephalic cyst, in axial and sagittal views, respectively

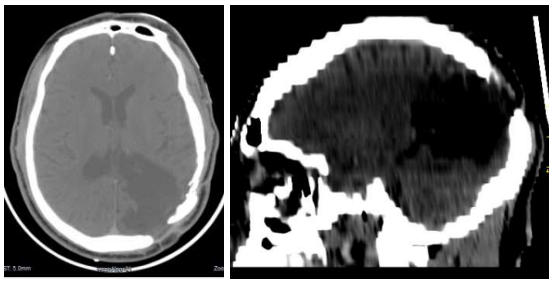
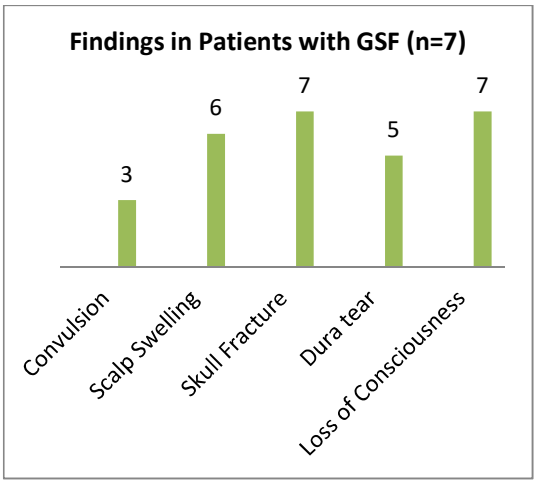


Figure 5. Findings in GSF



Intra-operative Findings

Four patients (Cases 1, 2, 3 & 5) were managed operatively. Case 2 had cysto-peritoneal shunt, while the others had craniotomy and duroplasty. All the cases had soft scalp swelling and skull fracture with wide defects. All operatively treated patients (Cases1, 3 & 5) had contused brain tissue herniating epidurally and sub-galeally. Dura was torn in all patients and retracted away from fracture line. The cystic overlying swelling contained mixture of CSF and contused herniated brain tissue. No haematoma collection was noticed in the swellings. Underlying and adjacent intra-cerebral and minimal epidural haematoma were however noted.

DISCUSSION

In 1816, Howship described a 9-month old child in whom an enlarging defect in his parietal bone developed after an injury. The defect was apparent within 2weeks after the injury and never resolved. The boy was re-examined when he was 4years old and "*the opening in the cranium remained undiminished; upon laying the hand on the part, the pulsations of the brain were felt strong and distinct*".[4]This description resulted in the pathological entity known as growing skull fracture (GSF).

It is a rare complication of skull fractures which occurs predominantly in children and rarely, in adults. It accounts for only 0.05-1.6% of skull fractures and only 1% of paediatric skull fractures, with 90% occurring before the age of 3 with a mean age <1year.[3,5,6] In a particular series, only 7.4% of patients were >3years.[7] The high incidence among the paediatric age group has been attributed to the rapid brain and skull growth in first 2years of life with a tight adherence of dura to bone making it vulnerable to tear. In addition, the skull is thinner, less stiff and more deformable, and in deforming can more readily tear the dura.[7]

Lack of consensus on the cause is reflected by the different names used to describe it: traumatic ventricular cysts, cranio-cerebral erosions, cranial malacia, post-traumatic leptomeningeal cysts, traumatic meningocele, cephalhydrocele, spuria, among others. Males are more affected than females, and most GSF occur in the parietal region, but have also been reported at the basi-occiput and orbital roof.[8] Five (71.4%) of our patients had involvement of the parietal region, making it the most common site in this series, as well. The interval between the time of head injury and the diagnosis of GSF has been reported to range from 1day to 8years.[4,5,7] The brain extrusion may be present shortly after diastatic linear fracture in neonates and young infants resulting in focal dilation of the lateral ventricle near the growing fracture.[12]

Though no specific or unique mechanism of injury predisposes patients to the development of a growing fracture, Xiaoyu Wang, *et al*, proposed that children under 3years with cephalheamatoma, underlying brain damage or bone diastasis ≥4mm on CT, and seizures immediately after the injury, are more susceptible to developing GSF.[4,9]

Road traffic accident (RTA), fall from height, cranial operation (craniosynostosis surgery), difficult vacuum delivery, assault and child abuse are aetiological factors implicated, with fall from heights being the most common.[7,9] Trauma from RTA was overwhelmingly the most common cause, accounting for 85.7% of cases, in among our patients, underscoring the burden of trauma in the developing world. Interestingly, all those involved were passengers on motorcycles.

The exact mechanism that leads to GSF has not been fully understood, but theories proposed include:

1. Ball Valve Theory described by Dyke, and supported by Taveras and Ransohoff in 1953. It is thought to arise from expanding pouch of arachnoid passing through the torn dura and skull fracture forming cysts which act as a one-way valve that traps CSF. This causes progressive pressure erosion of the fractured edges leading to enlargement of the fracture.
2. In the Water Hammer Theory, growth of the brain, produces pulsating, spreading tensile pressure forces on the edges of an unrepaired dural laceration with enlargement of the skull defect.

Presentation could be as an emergency or on an elective basis in which the patient would have had a previous trauma which was managed conservatively. The similarity of cephalhaematoma to the cystic swelling of GSF could explain the initial conservative approach to management hence, the need to fully examine any such swelling for underlying skull defect.

Three types of GSF have been described: Type I - Leptomeningeal cyst, Type II - Damaged and gliotic brain, and Type III – Porencephalic cyst. Case 1 of our series had Type I, while case 4 had a Type III GSF, which is also referred to as porencephalic cyst, but he had no scalp swelling. Other cases were type II GSF.

Skull radiography and CT are the two most useful investigations for the diagnosis and monitoring of GSF, however, MRI has greater sensitivity in detecting dural tears and is preferable where feasible.[5,13] Distinguishing leptomeningeal cysts from other pathologies is easier with MRI. On both T1- and T2-weighted MRI, GSF is isointense to CSF.[14] Skull x-rays may show widening of fracture, smooth edged ossification defect, scalloping of edges, and occasionally, sclerotic margins. With the exception of case 7 who was lost to follow-up, all our patients had brain CT scan; and none had MRI.

Surgery is the mainstay of treatment. Supportive therapy (as appropriate) should be instituted viz. anticonvulsants, antibiotics, analgesics, fluids, oxygen and correction of anaemia. All our patients who were operated on presented with severe anaemia which in most cases required repeated blood transfusions. The high vascularity of the scalp could make exsanguinating haemorrhage concealed and so, close attention should be paid to this possibility.

The principles of surgery include a large incision needed beyond margins of skull defect, defining the bony defect, reduction or resection of herniated brain, identification of the dural margin, tension-free water tight repair of the dural defect (key step) and covering of the missing bone (cranioplasty). There is no room for minimally invasive surgery.

In cases 2, 3, 5 and 6, the brain tissue was resected, whereas in case 1, attempts at reduction were successful and there were no neurological sequelae recorded in the latter. In all cases, the dura was found to be retracted from the bony edges making durorrhaphy impracticable. Duroplasty using pericranium had to be adopted in all cases. Pericranium is readily available, easy to harvest, indigenous and cost-effective. Bearing this in mind, initial skin flap should be raised meticulously to avoid damage to the pericranium.

In case 1, bone coverage was achieved by bi-valving calvarial graft, which served a double purpose - first as a means to expose the dural edge, and secondly, as bone cover after splitting the calvarial graft. Bony coverage was not achieved in other cases, and parents were counseled for possible cranioplasty at a later date. Rib grafts have also been successfully used to repair bony defect where calvarial graft is not feasible.[15]

We found no surgical indication for the adult patients with porencephalic cyst, and thus managed him conservatively. Similar conservative management of similar cases have been reported for incidental finding with no significant clinical consequence.[8]

Case 1 recovered completely with no neurological deficit, whereas cases 3, 5 and 6 had transient contralateral paresis which resolved completely before discharge. Case 2 had a complete resolution of the cystic swelling after shunting but, bony defect still remained the same 5months post-op, with residual contralateral paresis, ipsilateral visual loss and ptosis which points to the severity of the impact sustained.

CONCLUSION

It is imperative that post-traumatic scalp swellings be, thoroughly examined for underlying skull defect, and all diastatic skull fractures be followed up for possibility of GSF. In the paediatric age group, anaemia must be sought for and corrected. Surgical treatment modalities are very effective with good outcome and should not be delayed when indicated. This pathological sequela of skull fracture should be remembered and looked for even in adults with linear skull fractures.

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