

GROWING SKULL FRACTURES AFTER CRANIOSYNOSTOSIS REPAIR

Sanjay M. Khaladkar,¹ Poorvi Sharma,¹ Parth Sharma,² Shikha Bhadoriya¹

¹ Department of Radiodiagnosis, Dr. D.Y. Patil Medical College and Research Institute, Dr. D.Y. Patil Vidyapeeth Pimpri, Pune, India.

² Department of Radiodiagnosis, Ruxmaniben Deepchand Gardi Medical College, Pune, India.

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ABSTRACT

Growing skull fracture (GSF) is also termed as cranio-cerebral erosions, leptomeningeal cyst, cephalohydrocoele, traumatic meningocele, meningocele spuria and pseudoencephalocele. It occurs in 1% of linear skull fractures. 90% of patients are below the age of 3 years and 50% of patients are below the age of 1 year. The condition is rare after the age of 3 years. Growing skull fractures commonly involved calvarial bones. Etiologies include fall, vehicular accident and child abuse. Rare causes are corrective surgeries for craniosynostosis and following difficult vacuum extraction and after burr hole placement. An important factor in its pathogenesis is a skull fracture with its dural tear and entrapment of arachnoid membrane and brain tissue within fracture margin. Till now only 9 reports of GSF after corrective surgery for craniosynostosis have been reported. We report a case of an 11 - month old male child, a known case of Crouzen's syndrome who was operated previously for craniosynostosis by bifrontal craniotomy, fronto-orbital advancement and right frontal ventriculo-peritoneal (VP) Shunting. He presented with multiple extracranial lobulated soft tissue swellings. Computed tomography of the brain showed dilatation of both lateral ventricles. Cranial vault was deformed in shape due to craniosynostosis. Copper beaten skull suggestive of raised intracranial tension was noted. Multiple defects were noted in the cranial vault in bilateral fronto-parietal regions through which herniation of CSF filled structures were noted suggestive of pseudomeningoceles.

Keywords: Craniosynostosis, growing skull fracture, porencephalic cyst, dural defect, pseudomeningocele, leptomeningeal cyst.

Introduction

Growing skull fracture (GSF) is also termed as cranio-cerebral erosions. It occurs in 1% of linear skull fractures. It is sustained under 3 years of age. It can present many years later with headache, hemiparesis and seizures. There is progressive enlargement of the fracture line.¹ It is termed leptomeningeal cyst due to frequent association cystic mass filled with CSF.² Its exact etiopathological process is not known. However, dural tear is the most important factor in its pathogenesis. Four essential features are observed in the growing fracture of skull - a) Skull fracture in infancy or early childhood, b) dural tear at the time

of fracture, c) Brain injury underlying the fracture, and d) progressive enlargement of fracture with resultant increase in size of cranial defects.³

It was first described by Howship in 1816. 90% of patients are below the age of 3 years and 50% of patients are below the age of 1 year. The condition is rare after the age of 3 years. Growing skull fractures commonly involved calvarial bones. Rarely orbital roof and basiocciput can be involved.⁴

Case Report

11- month old male child presented with vomiting and difficulty in breathing and decreased activity. He

Correspondence : Dr. Sanjay M. Khaladkar
Department of Radiodiagnosis,
Dr. D.Y. Patil Medical College and Research Institute,
Dr. D.Y. Patil Vidyapeeth Pimpri, Pune, India.
Email: drsanjaymkhaladkar@gmail.com

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was a known case of syndromic craniosynostosis - Cruozen's syndrome with VP shunt in situ. The child had a full term normal delivery, cried immediately after birth. There was no history of neonatal ICU admission. Antenatal ultrasonography (USG) showed abnormal shape of the head at 7 months.

There was prior history of bifrontal craniotomy, fronto-orbital advancement followed by right frontal ventriculoperitoneal (VP) shunt and repositioning of right VP shunt. The child now presented with multiple swellings in skull and increased head size. Post-operative plain CT of the brain showed moderate dilatation of both lateral ventricles and 3rd ventricle with effacement of basal cisterns and sulcal spaces which had increased as compared to preoperative CT scan. There were changes of encephalomalacia in bilateral fronto-parietal region with ex- vacuo dilatation of bilateral frontal horns.

3D CT showed compound craniosynostosis with premature closure of bilateral coronal sutures, partial fusion of bilateral lambdoid sutures and sagittal suture. The Copper beaten appearance of a skull and bilateral harlequin eyes were observed.

CT Brain (Fig. 1-7) showed dilatation of both lateral ventricles. Bilateral frontal horns were more dilated and pulled anteriorly, 3rd ventricle showed mild dilatation, 4th ventricle was normal in size. VP shunt

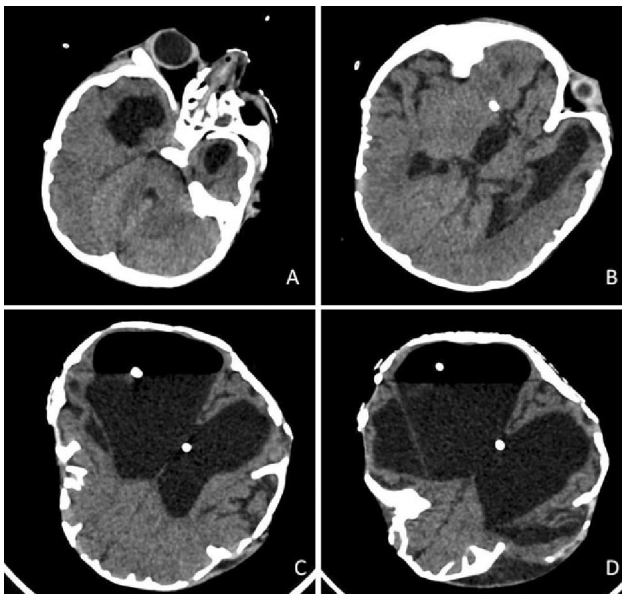


Figure 1: Plain CT Brain axial showed dilatation of both lateral ventricles. Bilateral frontal horns were more dilated and pulled anteriorly. Cranial vault was deformed in shape due to craniosynostosis. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region.

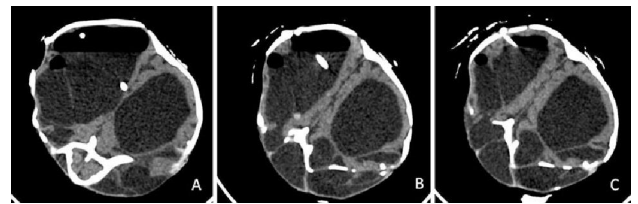


Figure 2: Plain CT Brain axial showed dilatation of both lateral ventricles. Cranial vault was deformed in shape due to craniosynostosis. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region with multiple pseudomeningoencephaloceles extending through these bony defects.

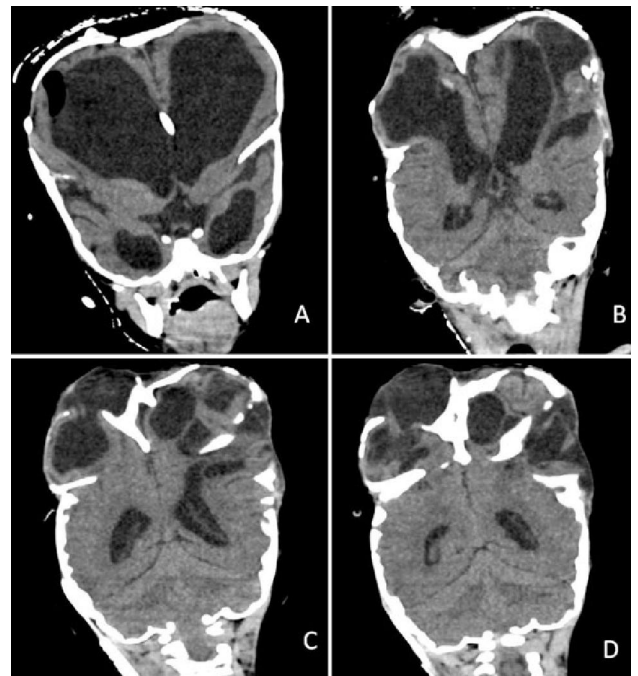


Figure 3: Plain CT Brain coronal sections showed dilatation of both lateral ventricles. Cranial vault was deformed in shape due to craniosynostosis. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region with multiple pseudomeningoencephaloceles extending through these bony defects.

was noted in situ with its tip in right frontal horn. Cranial vault was deformed in shape due to craniosynostosis. Scalloping was noted along an inner table of cranial vault on either side in fronto-temporo-parieto-occipital region - suggestive of Copper beaten skull - suggestive of raised intracranial tension (ICT). Multiple defects were noted in the cranial vault in bilateral fronto-parietal region. Largest defect in the right parietal bone measured 2.4 cm and large defect in the left parietal bone measured 2.2 cm. Well defined fluid density lesions (CT value 0-5 HU) were noted extending through these bony defects in

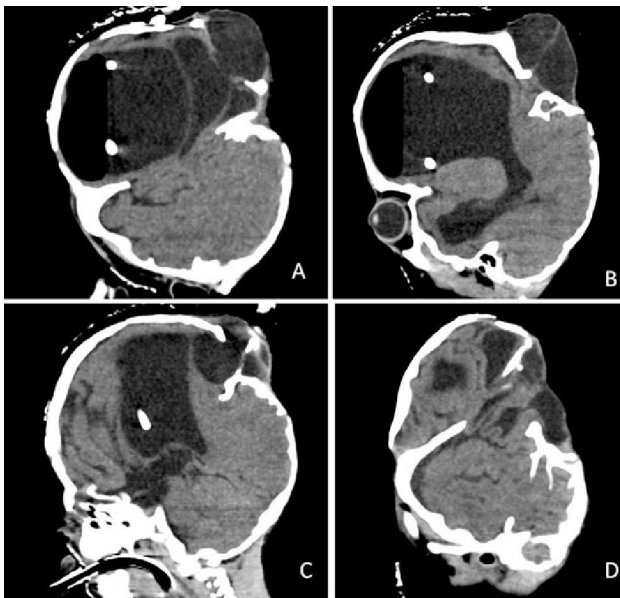


Figure 4: Plain CT Brain sagittal sections - showed dilatation of both lateral ventricles. Cranial vault was deformed in shape due to craniosynostosis. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region with multiple pseudomeningoencephaloceles extending through these bony defects.



Figure 5: CT scan of brain coronal at bone windows -Cranial vault was deformed in shape due to craniosynostosis with copper beaten skull. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region.

extracranial soft lesions with resultant extracranial lobulated soft tissue swellings. They measured approx. 54 x 37 x 49 mm on right side and 82 x 47 x 82 mm on the left side. These showed multiple tiny septations.

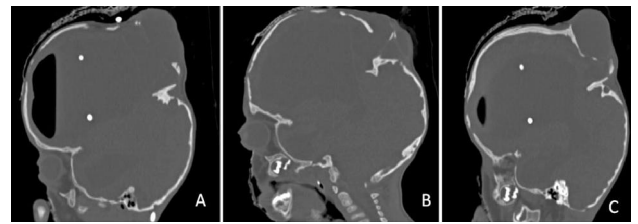


Figure 6: CT scan of brain sagittal at bone windows - Cranial vault was deformed in shape due to craniosynostosis with copper beaten skull. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region.



Figure 7: 3D CT - Cranial vault was deformed in shape due to craniosynostosis with copper beaten skull. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region.

There was high suspicion of the communication with the occipital horn and body of both lateral ventricles. There was thinning of brain parenchyma in bilateral fronto-parietal region. There was a fusion of coronal, lambdoid and sagittal suture - suggestive of craniosynostosis - Cruozen's syndrome. Diagnosis of growing fracture with dural defects with pseudomeningocele and porencephalic cyst was made. CT Ventriculography (Fig. 8-10) was done by injecting non-ionic contrast through the drain in right frontal horn. The fluid density lesions extending through bony defects were opacified with contrast on Ventriculography suggestive of communication of porencephalic cysts with the ventricles. Transverse diameter of bifrontal region was widened as compared to bilateral parieto-occipital region. Diagnosis of growing

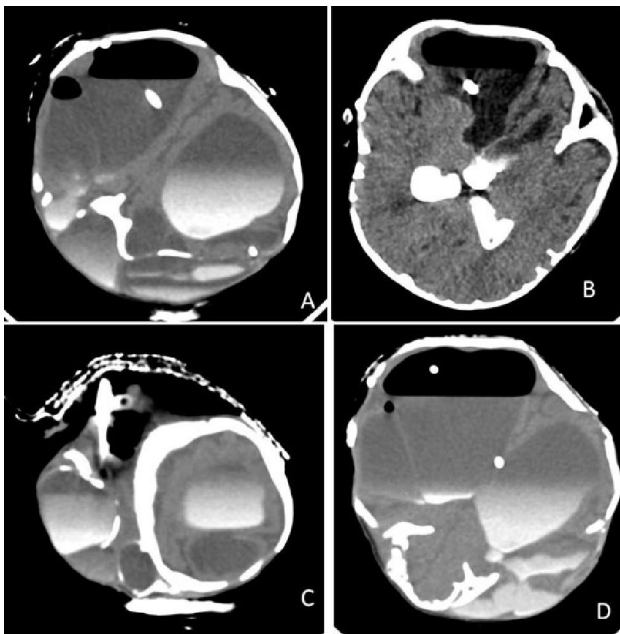


Figure 8: CT Ventriculography axial sections - the fluid density lesions extending through bony defects were opacified with contrast on Ventriculography suggestive of communication of porencephalic cysts with the ventricles.

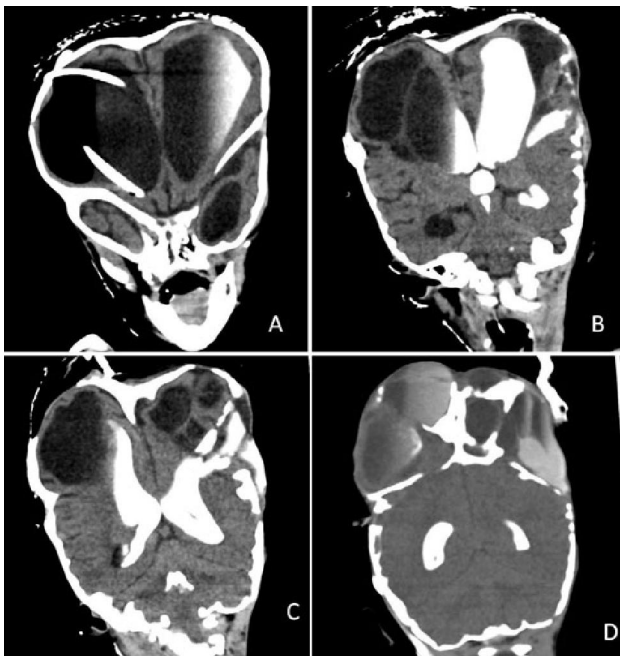


Figure 9: CT Ventriculography coronal sections - the fluid density lesions extending through bony defects were opacified with contrast on Ventriculography suggestive of communication of porencephalic cysts with the ventricles.

fractures with dural defects with herniation of posterior portion of both lateral ventricles through bony defects was given.

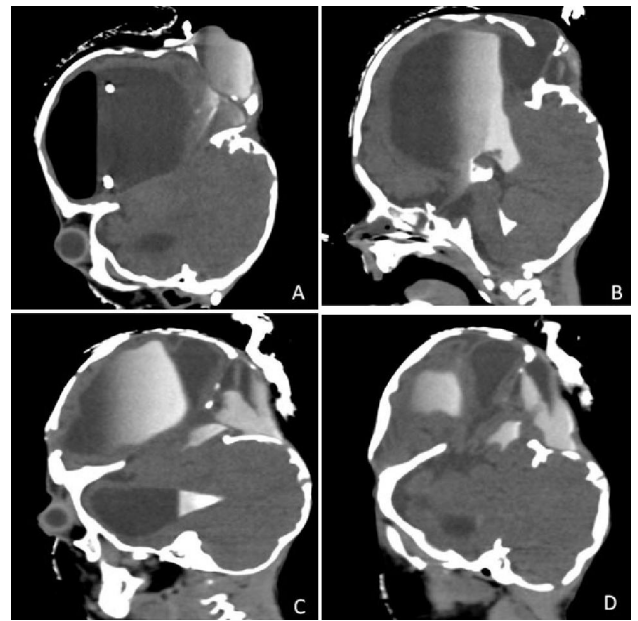


Figure 10: CT Ventriculography sagittal sections - the fluid density lesions extending through bony defects were opacified with contrast on Ventriculography suggestive of communication of porencephalic cysts with the ventricles.

MRI Brain (Fig. 11-15) showed marked thinning of brain parenchymal in bi - frontal region superior to dilated lateral ventricles. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region with herniation of CSF filled structures - suggestive of meningoencephaloceles.

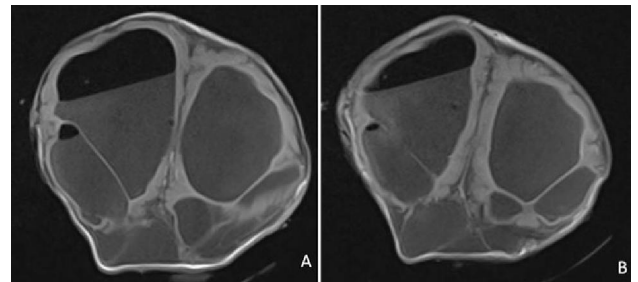


Figure 11: MRI Brain axial T1WI showed marked thinning of brain parenchymal in bi - frontal region superior to dilated lateral ventricles. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region with herniation of CSF filled structures - suggestive of meningoencephaloceles.

Discussion

Etiologies of GSF include fall, vehicular accident and child abuse. Rare causes are corrective surgeries for craniosynostosis and following difficult vacuum extraction and after burr hole placement.⁵ An important factor in its pathogenesis is a skull fracture with

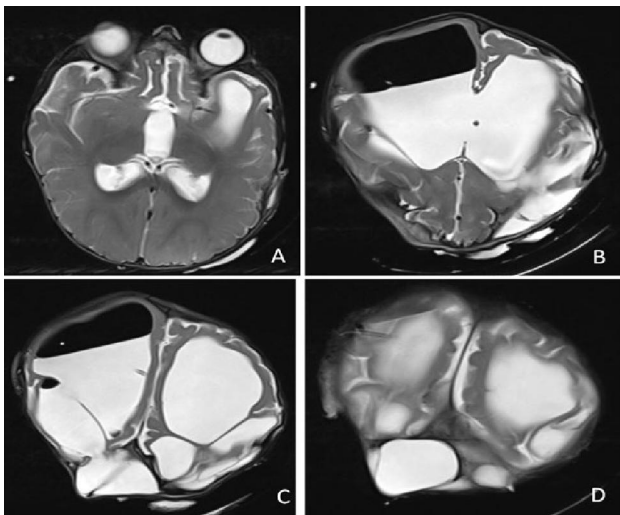


Figure 12: MRI Brain axial T2WI showed dilatation of both lateral ventricles. Multiple defects were noted in the cranial vault in bilateral fronto-parietal regions with herniation of CSF filled structures - suggestive of meningoencephaloceles.

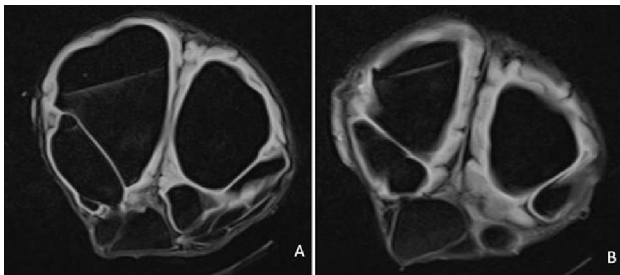


Figure 13: MRI Brain axial FLAIR showed dilatation of both lateral ventricles. Multiple defects were noted in the cranial vault in bilateral fronto-parietal regions with herniation of CSF filled structures - suggestive of meningoencephaloceles.

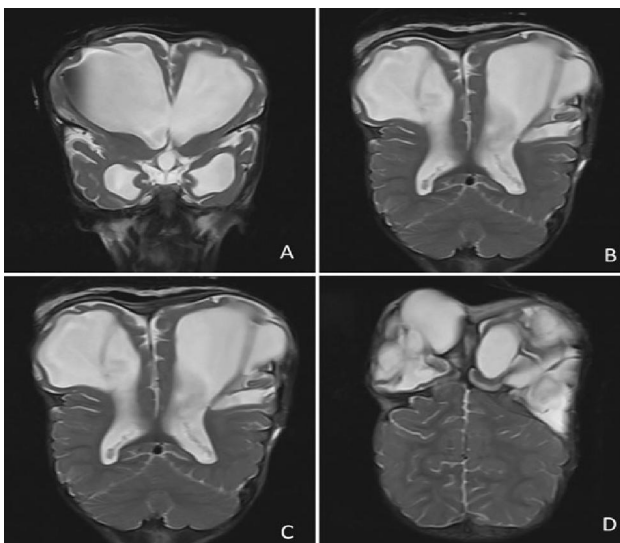


Figure 14: MRI Brain coronal T2WI showed dilatation of both lateral ventricles. Multiple defects were noted in the cranial vault in bilateral fronto-parietal regions with herniation of CSF filled structures - suggestive of meningoencephaloceles.

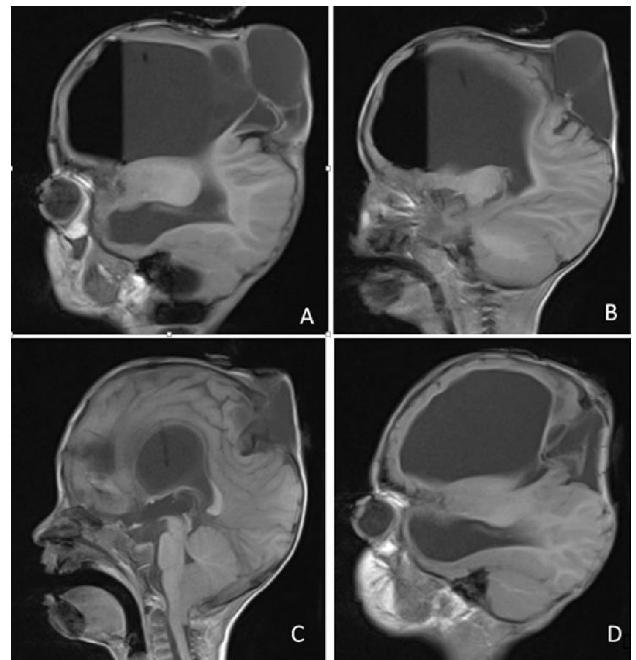


Figure 15: MRI Brain sagittal T1WI showed dilatation of both lateral ventricles. Multiple defects were noted in the cranial vault in bilateral fronto-parietal region with herniation of CSF filled structures - suggestive of meningoencephaloceles.

its dural tear and entrapment of arachnoid membrane and brain tissue within fracture margin. Higher incidence of GSF in infancy and early childhood occurs due to the rapid growth of the brain and skull in first 2 year of life when the dura matter is tightly adherent to the bone and hence easily torn when the skull is fractured. The other hypothesis is that the skull is less stiff, thinner and deformable.

Due to these factors, any deformity in skull results in underlying dural tear.⁶ Progression of Growing skull fractures occurs in 3 stages:- **STAGE I**- The prephase of growing skull fracture - This stage is from the time of injury till just before the enlargement of the fracture. At this stage, skull fracture with dural tear and herniation of arachnoid membrane and brain tissue occur through the fracture. **STAGE II**- Early phase of growing skull fracture - It occurs from the initial fracture enlargement to 2 month after the beginning of enlargement. At this stage, the bony defect is more and the skull deformity and neurological deficit are mild. If the growing skull fracture is diagnosed and treated at this stage, the prognosis is good. **STAGE III** - Late phase of growing skull fracture - It begins 2 months after initial enlargement of fracture. The bone defect becomes larger in size. The skull deformity

and neurological deficit become severe if untreated.⁷ Growing skull fracture are divided into 3 types based on CT appearance - TYPE I - leptomeningeal cyst with GSF herniating through the skull defect into the subgaleal space. TYPE II- associated with brain damage or gliosis. TYPE III- associated with porencephalic cyst.⁸ Increase in severity of neurological deficit is observed with increase in the order of the defect.⁴

CT is useful in detecting fracture. MRI can detect a dural tear immediately following the head injury. Hence the timely correction can prevent growth of fracture. Treatment of the GSF is surgical and includes reduction of herniated cerebral tissue, repair of dural laceration or cranioplasty shunt. Surgery can be done to decompress the cyst and treat localized ventricular dilatation.⁴

Other terminologies used for growing skull fracture are leptomeningeal cyst, cerebro-cranial erosion, cephalohydrocele, traumatic meningocele, meningocele spuria and pseudoencephalocele.

Risk factors for occurrence of growing skull fracture after craniofacial procedure are - 1) Hydrocephaly or signs of increase intracranial pressure due to limited lymph circulation, signs of lymph diapedesis and dural ectasia surrounding the optic nerve, Arnold Chiari malformation I and asymmetry of the septum pellucidum. 2) Bilateral or unilateral coronal craniosynostosis due to the displacement of lateral sphenoid wing and middle cranial fossa. 3) Complex craniosynostosis and cranio-facial syndrome affecting more than one suture. Cranioplasty becomes challenging in these cases due to the impression on the inner cranium due to increased intracranial pressure. 4) Repeated surgical intervention which increased risk of dural leaks due to scarring and dural adhesions. 5) Endoscopic craniosynostosis repair. Recognition of intra-operative dural injury is difficult as it is a minimally invasive procedure.⁹

Till now only 9 reports of GSF after corrective surgery for craniosynostosis have been reported.⁹ In syndromic cranio-facial cases affecting the skull base (Apert, Crouzen, Pfeilfer, Seather- Chotzen syndromes) the middle cranial fossa is located more anteriorly than in non - syndromic craniosynostosis. The lateral sphenoid wings are also displaced anteriorly. Hence unrecognized dural laceration can occur during corrective procedures due to limited field of view.

Mechanism of Growing skull fracture (Fig. 16) There are two requirements for expanding dural defect irrespective of the mechanism of injury - a) The dura has to be disrupted. b) There must be an outward driving force (like normal growing brain, diffuse cerebral edema, hydrocephalus, brain tumor). The cerebral hernia continues to enlarge until the dura is repaired or until the outward driving force is diminished (by cessation of brain growth). The radial forces of the growing brain are normal counteracted by an intact dural envelope. If the dura is defective, the lesser-restraining forces offered by scalp overlying the defect and viscoelastic properties of infant brain, the brain herniation can occur. Normal tensile forces within the dura are re-established after the repair of dural defect thus confining the brain and allowing uniform distribution of radially active forces of the expanding brain.¹⁰ The growing skull fracture occurs due to expanding dural defect. The dura of a normal infant grows, this characteristic applies equally to intact dura and the dura at the edge of laceration. The dura grows in the direction of tensile force, similarly the edge of the lacerated dura continues to grow in a direction that is parallel to and not centripetal to the edge of the defect. This results in progressive enlargement of the defect.

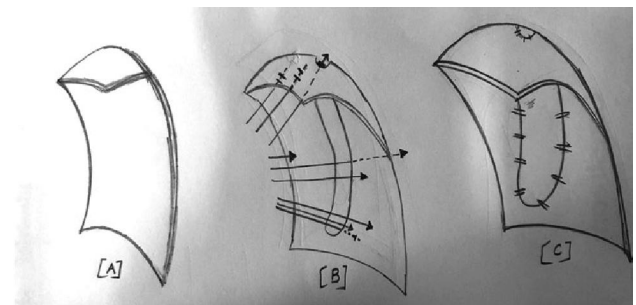


Figure 16: Diagram of radial forces acting on the expanding defect. (A) Intact dura - forces approximately evenly distributed against dura. (B) Lacerated dura - the radial forces in the region of the defect unopposed by the restraining dural envelope. (C) Dural defect repaired - radial forces again evenly distributed.

Conclusion

Corrective surgeries for craniosynostosis is a rare cause of growing skull fractures. These occur due to dural defects which progressively enlarge with growth

of brain and raised intracranial pressure. There timely detection is of utmost importance to prevent their growth.

Conflict of Interest: None

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