Case report:

A Case of Non-syndromic Craniosynostosis

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Abstract:
Craniosynostosis refers to skull deformities secondary to the premature closure of cranial suture. Isolated or multiple sutures craniosynostosis is more common than syndromic craniosynostosis. Deformities with synostosis are stigmatizing, and this provides a strong aesthetic indication for surgical correction in the non-syndromic group of patients. We present a case of non-syndromic sagittal synostosis in a ten months old patient underwent open surgical repair for skull deformity.

Keywords: Non-syndromic craniosynostosis, sagittal synostosis.

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Introduction:

Craniosynostosis was described as the premature closure of cranial suture leading to skull deformities by Sommerring in 1791.[1] Virchow observed in 1851 that skull growth perpendicular to the fused suture is inhibited by premature fusion.[2] Compensatory growth of adjacent sutures with premature fusion affects the entire skull.[3] Craniosynostosis requires correction surgery that performed addressing the whole skull and not just the fused suture. Sagittal suture fusion has an estimated incidence of 1 in 2000, making it the commonest suture involved.[4] Non-syndromic craniosynostosis is more common than syndromic craniosynostosis; up to 85%.[5] Approximately 40-60% of all craniosynostosis cases are non-syndromic sagittal craniosynostosis; making it the most frequent variant leading to scaphocephaly.[6]
Clinical diagnosis is adequate in most cases; however, confirmation with imaging such as X-rays, computed tomography (CT), and three-dimensional CT reconstruction is effective in delineating skull base deformity. Imaging offers added benefit during surgical planning. Common indications for craniosynostosis surgery are increased intracranial pressure and cosmetic deformity. Deformities with synostosis are stigmatizing, and this provides a robust aesthetic indication for surgical correction. We present a case of a non-syndromic sagittal craniosynostosis patient underwent open surgical repair for skull deformity.

Case Report:

A ten months old boy was referred to the craniofacial plastic team due to the elongated shape of his skull. He was born full-term via normal vaginal delivery with an uneventful pregnancy. The child was non-syndromic and did not have any medical or surgical problems. He has an eight-year-old elder brother who is well. There was no similar history among other relatives. According to parents, the child had typical developmental milestones.
On clinical examination, sagittal suture ridging was palpable. The head was shortened in the bi-parietal diameter and elongated in the anterior-posterior diameter. The child was diagnosed

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clinically with scaphocephaly. Computerized tomography was performed, which showed sagittal suture fusion. Magnetic resonance imaging revealed impediment of lateral growth of the skull with continuous growing anteroposterior part of the skull (Figure:1). No dilated ventricles or other abnormality of the brain observed.

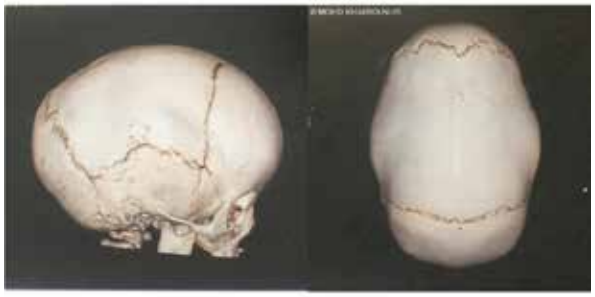

Figure 1: 3D – Reconstructed Computerized Tomography performed showing sagittal suture fusion and elongated AP diameter.

The child underwent modified Pi Procedure and cranial vault reshaping, a combined surgery with the pediatric neurosurgical team. A bi-coronal incision was performed. Hemostatic clips were placed to reduce blood loss. Subperiosteal plane dissection performed and extended laterally detaching the temporalis from the temporal bone. Anterior flap elevated down to the level nasofrontal and supraorbital rims. The supraorbital and supratrochlear nerves were identified and preserved. The posterior flap elevated subgaleal beyond the lambdoid sutures. A Pi (π) shaped, bi-frontal and bilateral parasagittal craniectomy of 1cm strip was performed with a cranial oscillating saw with removal up to lambdoid and coronal sutures (Figure 2). The frontal bone flap was bisected into half, reshaped and applied back as two bone grafts and attached to the frontal orbital bar with Ethilon® 1/0 sutures. Bilateral temporal bone was out-fractured to increase the skull width (Figure 3). Both frontal and occipital bones were brought towards the central axis, correcting the anterior-posterior dimensions. The total intracranial volume was maintained by increasing the width of the skull initially and shortening the length later. Drains left in place, and the skin was closed in layers. The child's recovery was uneventful and discharged well on the postoperative day-10. The patient is currently on regular follow up.

Figure 2: Bi-coronal skin incision (right). Pi (π) shaped Bi-frontal and bilateral parasagittal 1cm strip markings up to lambdoid and coronal sutures (left).

Figure 3: Frontal bone flap bisected into half, reshaped and applied back as two bone grafts and attached to the frontal orbital bar with non-absorbable sutures.

Figure 4: Postoperative 1-week image shows correction of frontal bossing and improvement of AP diameter.

Discussion:

Relevant discussion points about non-syndromic craniosynostosis (NSCS) includes development and pathogenesis of the disease, clinical evaluation, imaging modalities, pre-operative patient optimization, the surgical technique employed, post-surgical follow-up, and psychosocial issue of untreated cases.

Skull in the newborns consist of flat bones separated by six major cranial sutures and four fontanelles.[7] Dense fibrous tissue composition of sutures permits limited physiological movement such as expansion and compression that occurs

during birth.[8] Size of the cranium at birth is approximately 80% of adult size, and definitive size is achieved at three years of age. However, sutures and fontanelles close according to the various timeline from three months to mid-thirty years.[7]

The pathological process seen here is skull growth restriction parallel to the affected prematurely fused sutures. The fusion of sutures leads to the limitation of underlying brain growth due to loss of suture accommodation and compensatory overgrowth at non-fused suture areas leading to distortion.[9] NSCS has various types of head shape dysmorphic characteristic. Involvement of different regions and numbers of sutures leads to a variety of clinical findings.[10] In descending order, sagittal suture is the commonest affected, followed by coronal, metopic and lambdoid sutures for single suture synostosis.[7]

The pathogenesis of craniosynostosis is complex and unclear. Multifactorial theories proposed include genetic mutations, intrinsic bone abnormalities and environmental factors.[8] Craniosynostosis has been associated with metabolic conditions such as hypophosphatemia and rickets. The main factor that leads to craniosynostosis is the constraint in fetal growth which are seen in nulliparous mothers and multiple pregnancies. Multiple prenatal factors that are taken into consideration include maternal smoking, teratogen exposure, maternal consumption of anti-epileptics or excessive antacids, low birth weight and pre-term delivery.[11] There is no single genetic cause identified; however, the gene encoding for fibroblast growth-factor is frequently mutated in syndromic craniosynostosis. Abnormal maturation of suture and cranial malformation are evident due to defective signaling and tissue interaction.[9,12]

The most typical clinical presentation of NSCS during the first year of life is an unusual shape of the head. Head anomaly includes being flattened and broad (brachycephaly), triangular front (trigonocephaly), skewed (plagiocephaly), or long and narrow (scaphocephaly or dolichocephaly). Skull should be palpated for ridging, mobility and the presence of fontanelles. Quantitative cranial anthropometric measurements are taken, and special attention is given to look for congenital anomalies. Major functional complications associated with this disorder are often irreversible. They comprise of limited brain growth, visual impairment, intracranial hypertension and neuropsychiatric disorder.[8,11] Therefore, the examination must be performed thoroughly, and other craniofacial disorders should be ruled out.

Patients must be managed in a specialized pediatric craniofacial centre wherever possible. Surgery is planned soon after diagnosis. Timing for surgery advocated from the first few weeks of birth till nine months of age.[8] Staged or secondary surgeries are performed in severely affected patients that need correction of residual deformities. Management of NSCS focuses on prevention and correction of skull deformity and stabilizing intracranial pressure if elevated. Attention is also given towards eye protection, airway, infant feeding and optimal oral health. Unlocking and reshaping of bone optimize correction and reduces intracranial pressure. Functional and aesthetic reasons are also taken into consideration.[11] Some authors proposed a minimally invasive technique which reduces operative morbidity and shorter hospital stay; however, there are diverse techniques available.[7] Recent advances in pediatric anesthesia and biomaterial developments allows bone substitution with hydroxyapatite cement and usage of resorbable fixation systems.[8] Recommended operative technique for NSCS includes (1) Open calvarial reconstruction, (2) Cranial distraction osteogenesis, (3) Endoscopic suture release, (4) Strip craniectomy with spring implantation, and (5) Strip craniectomy with the use of a postoperative moulding helmet.[11]

Surgery of choice performed for sagittal craniosynostosis is the Pi procedure, an open calvarial reconstruction method named after the shape of the bone removed. In this technique, the bilateral coronal, lambdoid and the sagittal sutures are removed initially. The sagittal suture is used as a strut to maintain the expanded parietal bone, which was out-fractured to increase the skull width. Frontal bossing and anterior-posterior dimension adjustment addressed before securing the frontal and occipital bones to the parietal bones.

Patient follow-up after reparative surgery should continue until skeletal maturity.[10] Follow up period varies according to the severity of deformity; however, they are reviewed at least annually up to adolescence. Signs and symptoms of increased intracranial pressure (e.g., nausea and vomiting, irritability, headache, visual disturbances, seizures, developmental delay, and declining academic performance) and aesthetic results are reviewed during follow-ups.[8]

Untreated NSCS results in aggravated craniofacial deformities. Visible facial differences with

cognitive difficulties, impairment of vision, language or behavior leads to difficult peer interaction and psychosocial issues.[11] This resulted in poor health-related quality of life. Care-giver, including parents, undergoes stress and are psychologically affected by their children's condition. They have to endure having a child with a congenital anomaly, frequent hospital visits, surgeries, financial support and providing specialized care. These factors will affect the caregiver's behavior and the child's psychosocial adaptation.

Conclusion:

Non-syndromic sagittal craniosynostosis is a common condition managed by craniofacial plastic surgeons; however, there are wide discrepancies in the management options for it in terms of pre-operative evaluation, operative techniques, and peri-operative management. A competent multidisciplinary team should perform early diagnosis and comprehensive treatment. Management aimed at improving the patient's daily functionality and psychosocial well-being are imperative. The modified Pi technique is an effective method for immediate correction of sagittal synostosis. It addresses all aspect of deformity, avoids further manipulations such as moulding helmets and produces a rounder cranial vault.

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