Open suturectomy management in craniosynostosis of bilateral coronal and metopic suture

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Abstract

Craniosynostosis is a cranial deformation that is characterized by the premature fusion of one or more of the cranial sutures. Synostosis of multiple sutures is a rare case and can be treated with open suturectomy procedure which is one of the surgical management by removing the fused suture with the purpose to allow the constricted area to expand with the growing brain. We present a case of a 2-month-old male infant with abnormal head shape since birth with a non-contrast 3D computed tomography (CT) scan of the head confirmed closure of the bilateral coronal and metopic suture. Open suturectomy was performed with no post operative complications. Open suturectomy technique can be performed in the management of multisutural craniosynostosis as indicated. The diagnosis of which suture are affected, the timing of surgery, and the prevention of surgical complication such as excessive blood loss are the important factors need to be considered.

Keywords: craniosynostosis, suturectomy, coronal, metopic

Introduction

Craniosynostosis is a craniofacial malformation that is characterized by the premature fusion of one or more of the cranial sutures that evolve during embryonic life and appear at birth.¹⁻⁴ Its incidence is estimated to be 1 in 2,000-2,500 live births.¹ The skull and the growing brain are restrained under the premature fusion of the cranial suture, leading to a compensatory expansion into regions of the skull that are not affected.⁵ This condition also will result in an abnormal head shape and increase intracranial pressure in one-third of all cases that may have a mental mal-development.⁶ Craniosynostosis is classified as isolated (fusion of only one suture) or complex (more than one suture fused).⁷ Isolated fusions are named by the involved suture, and the cranial shape takes on a characteristic dysmorphism. In 1851, Virchow⁸ described in his paper about the growth patterns of craniosynostosis, which is known as the Virchow's law. Virchow's law states that in craniosynostosis the skull growth is restricted to a plane perpendicular to the prematurely fused suture and is enhanced in a plane parallel to it. The shape of the head may become more complex when more than one sutures are involved. Craniosynostosis is also classified as syndromic or non-syndromic. Syndromic craniosynostosis is defined as fusion of multiple sutures with associated extracranial extracranial anomalies.⁷ The frequency of syndromic craniosynostosis was reported
between 0.4 and 1.0 per 1000 live births, which is less frequent than non-syndromic craniosynostosis.5

Craniosynostosis of more than one suture is a rare case and can be treated with open suturectomy procedure which is one of the surgical management by removing the fused suture with the purpose to allow the constricted area to expand with the growing brain.

Case Report

We present a case of a 2-month-old male infant with abnormal head shape (brachycephaly) (Figure 1) and a history of seizure 1 month prior to admission. The patient was born full term by cesarean section with birth weight of 3.55 kg, birth length 50 cm, and occipitofrontal circumference 37 cm. The patient also has a history of neonatal asphyxia. Non-contrast 3D computed tomography (CT) scan of the head confirmed closure of the metopic and bilateral coronal suture (Figure 2).

Open suturectomy was undertaken at the age of 10 weeks. The patient’s head was placed in a neutral supine position. The head was shaved at the surgical area and was prepared using Chlorhexidine soap followed by disinfection using povidone iodine solution after a single bolus of antibiotics was given. The frontal cranial was exposed through bicoronal skin incision. Four burr holes were done, 1 at the keyhole for each side, and 2 others were done at the coronal suture for each side. A 2-3 cm wide suturectomy was performed along the closed bilateral coronal suture across the anterior fontanel and the metopic suture (Figure 3) using Kerrison rongeurs and bone rongeurs. The bleeding from the bone was controlled by wax. There were no complication after surgery.

Discussion

Craniosynostosis of bilateral coronal and metopic suture is a rare case. The percentage of affected sutures are sagittal (40-55%), coronal (20-25%), metopic (5-15%), and lambdoid (0-5%). Craniosynostosis with more than one suture affected, approximately 5-15% of cases.9

The diagnosis of craniosynostosis can be made based on physical examination, cranimetrics measuring, and observation of the skull deformity that accompanies each type of suture fusion. The radiological examination also has an important role in the confirmation of the diagnosis, the surgical planning, and even the postoperative follow-up.5,10 In infants, an important consideration is that the use of radiation or the need of sedation/anesthesia increase the problem and must be reduced to minimum to prevent from their adverse effects.10

In this case the patient has a brachycephaly, which is most commonly the result of premature synostosis of both coronal sutures. Bilateral coronal craniosynostosis causes a short (anteroposterior) and wide (transverse) head. The anterior fontanel may be open or closed3,9, which in this case the anterior fontanel of the patient is closed. We also performed a non-contrast 3D head CT scan for confirmation of the diagnosis and surgical planning, which reveals premature synostosis of both coronal and metopic sutures. The high-resolution CT scan with 3D
reconstruction is considered the gold standard in the radiological examination of craniosynostosis and must be carefully evaluated due to the radiation effect. Other radiological examination that can be used such as x-ray, ultrasound and magnetic resonance imaging (MRI). X-rays are a fast and low-radiation diagnostic tool with a high specificity but low sensitivity in detecting complex and minor suture synostoses. Ultrasounds can be useful because of the fastness of the execution, the absence of ionizing rays, and the possibility without using sedation, but very dependent on the operator’s skill and suitable only for certain age period. MRI eliminates the risks of ionizing radiation, but the requirement for the baby to keep the position in a long exam generally requires general anesthesia or sedation which become a major concern due to the related risks.10

Surgical management of craniosynostosis has developed significantly over the years. The first procedure reported for craniosynostosis is strip craniectomies or suturectomy.11 The purpose was to allow the constricted area to expand with the growing brain by removing the fused suture. Cranial vault reconstructions were introduced, to make a normal head shape with removing cranial bone, reshaping it, and stabilizing repositioned bony segment.12,13 This procedure has an excellent result but is a long-time surgical procedure with increased blood loss12 and prolonged time under anesthesia9. Less invasive surgical procedures have been developed with new techniques and applications to achieve the same outcomes and with an advantage of less blood loss and shorter scars12, but very much depends on the skills of the surgeon and the availability of the endoscopic instrument. The goal of surgical management in craniosynostosis is to correct the abnormal head shape of the cranial deformity and to prevent elevated intracranial pressure and other negative impacts on neurocognitive development and vision impairment.5,7,12,13

The timing of surgical management depends on several factors such as the type and severity of synostosis, surgical technique, comorbidities, and surgeon preference.5,6,12 In the literature the optimal timing of surgery is 3 to 6 months of age.6 Some authors also divided the timing of surgery into early surgical intervention which occurs between 2 to 3 months of age and later surgical intervention occurs 6 to 12 months old. Early intervention has advantage of early decompression of elevated intracranial pressure and increase the intracranial volume to allow the brain to grow.5,12 The procedure can be less extensive, less invasive and has the better outcome if the age of the patient younger at the time of operation. However, it is important to consider that in infants even small amount of blood loss may cause significant impact due to very small blood volume and low hematocrit level.14 Later intervention allows remodeling easier as the bones are better developed, easier to reconstruct and to stabilize, and also the child has a greater total blood volume.15

The management of craniosynostosis, requires a multidisciplinary team approach. Neurosurgeon will work together with anesthesiologist, pediatrician, geneticist and other specialists during the course of diagnosis, treatment, and follow-up.5

**Conclusion**

Surgical management of craniosynostosis has developed significantly over the years. However, open suturectomy technique can be performed in the management of craniosynostosis of more than one suture as indicated, especially if there are limited resources to perform other surgical techniques such as endoscopic suturectomy. Special attention is needed to prevent excessive blood loss that can lead to postoperative complications.

**Acknowledgment**

No acknowledgment

**References**


