



Craniosynostosis: Forehead Reconstruction Using Sagittal Bony Bar (Case Report)

ABSTRACT

A four years old boy was referred to Ibn-Sina Teaching hospital / Mosul city complaining from deterioration of vision 2 months ago, which was documented by an ophthalmological examination. A medical cranial imaging was taken; it showed fusion of sagittal, coronal and metopic sutures with diffused beaten copper appearance. The patient was admitted to the neurosurgical department, a measurement of intracranial pressure via lumbar puncture and cerebrospinal fluid manometry showed a 340 mm CSF reading which is very high. Surgery was planned by a neurosurgical and maxillofacial surgical team. Under general anesthesia, a bi-coronal flap was raised, incision and removal of a wide bony bar along the closed sagittal cranial suture and the skull bone was released by nibbling. The eye balls were separated from the orbital bones, the anterior part of orbital roof was removed; the flatted forehead was removed and reconstructed by using the bone that was removed from the sagittal area and fixed in its new place by titanium plates and stainless wire. The postoperative course was uneventful and the child's visual acuity returned to a normal condition dramatically. Follow up for three years showed normal cranial growth, normal vision and no medical complications that may be related to the surgical operation.

الخلاصة

تم إحالة طفل في الرابعة من عمره الى مستشفى ابن سينا التعليمي وكان يعاني من تدهور في حدة النظر منذ شهرين وهذا ما تم تأكيده من قبل اختصاصي العيون. لقد أظهرت الصور الطبية للجمجمة وجود التحام بين العظام المكونة للجمجمة وظهور اثار ضغط الدماغ على الجدار الداخلي للجمجمة. تم إدخال المريض إلى شعبة جراحة الجمجمة والأعصاب حيث تم قياس الضغط داخل الجمجمة بواسطة قياس السائل النخاعي الذي اخذ من العمود الفقري والذي اظهر زيادة كبيرة جدا، تم وضع خطة العمل الجراحي بواسطة الفريق الطبي من قبل جراحة الأعصاب وجراحة الوجه والفكين. حيث تم رفع شريط عريض من العظم الوسطي للجمجمة وازالة الالتصاقات الموجودة بين العظام، كما تم فصل كرة العين عن عظام المحجور وإزالة جزء من سقف محجر العين، ومن ثم إعادة تشكيل جبهة المريض باستخدام الشريط العظمي الذي تم إزالته من قمة الجمجمة و تثبيته بواسطة صفائح التيتانيوم والإسلاك الخاصة. أظهرت متابعة المريض بعد العملية عودة النظر إلى وضعه الطبيعي كما أظهرت المتابعة لمدة ثلاثة سنوات بنمو طبيعي للأعصاب البشرية والرؤية الممتازة وعدم وجود مضاعفات تتعلق بالعملية


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Key Words: Cranial deformity, congenital cranial defect, cranial syndrome.

INTRODUCTION

 Craniostenosis, the premature fusion of one or more of the cranial sutures, is the most common human congenital skull defect, with a prevalence of 333–476 per million births^(1, 2). A single cranial suture involvement is most commonly implicated, while the syndromic craniostenosis, with a more generalized pattern, is much less common⁽³⁾. There are over 100 described syndromic synostoses, the most common of which include Pfeiffer, Crouzon, Apert and Saethre-Chotzen^(4, 5). The majority of patients with craniostenosis have various skull alterations, which are severe in syndromic type and mainly considered to have arisen from compensatory growth of the skull after stenosis of some sutures and high intracranial pressure^(6, 7). In cases in which the compensation does not effectively provide enough space for the growing brain, craniostenosis would result in increased intracranial pressure leading possibly to visual impairment or an impairment of mental development⁽⁸⁾.

CASE PRESENTATION

A four years old boy was referred to Ibn-Sina Teaching Hospital / Mosul city complaining from deterioration of vision 2 months ago, which was documented by a weekly ophthalmological examination; it was reported that there was bilateral proptosis and papilledema in addition to gradual decrease of visual acuity (Figure 1). A skull radiographs, MRI and skull computed tomography (CT) scan were taken; those showed fusion of sagittal, coronal and metopic sutures with a diffused beaten copper appearance (Figure 2).

The patient previously sought treatment in neighboring countries but the surgical team there gave his family 20% mortality and 80% possibility of loss of vision during operation; therefore, the family refused doing the operation and returned to treat their boy in Iraq. The patient was admitted to the neurosurgical department in Ibn-Sina Teaching hospital. Measurement of intracranial pressure via lumbar puncture and cerebrospinal fluid monometry was carried out. The intracranial pressure was 340 mm CSF which is very high. Surgery was planned by a neurosurgical and maxillofacial surgical team.

Under general anesthesia and endotracheal intubation, a bicoronal incision design was done. Dissection of the scalp was made anteriorly down to the orbital rim and posteriorly to the lambdoid suture, two bur holes on each side of the anterior end of the sagittal suture and another two holes were made on each side of the posterior end of the sagittal suture; a cut lines with removal of the bony bar including the fused sagittal suture was done (Figure 3).



Figure 1

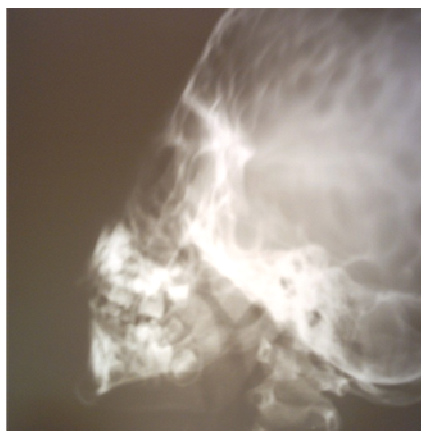


Figure 2



Figure 3

Then, the sides of the skull were released by nibbling of bone extending from each bur hole to the base of the skull, so that each parieto-temporal bone on each side becomes free. The eye balls were separated from the orbital margins, the supra orbital bony bar was removed, and then the frontal bone (forehead) which was narrow and flattened was removed. Retraction of the frontal dura with removal of apart (anterior) of the roof of the orbits was done; this gave more space to the orbital cavity. The forehead was reconstructed by application of the bony bar that had been removed earlier from the area of the sagittal suture and fixed anteriorly by titanium plates, and posteriorly by stainless steel wires. Eventually, the wound was closed in layers with drain application.

The surgical work took approximately six hours (Figure 4). The postoperative course was uneventful and dependent on ophthalmological examination of the child's visual acuity returning to normal condition dramatically. Follow up for three years with six months recall visits showed normal cranial growth, normal vision and no medical complications that may be related to surgical operation (Figure 5).

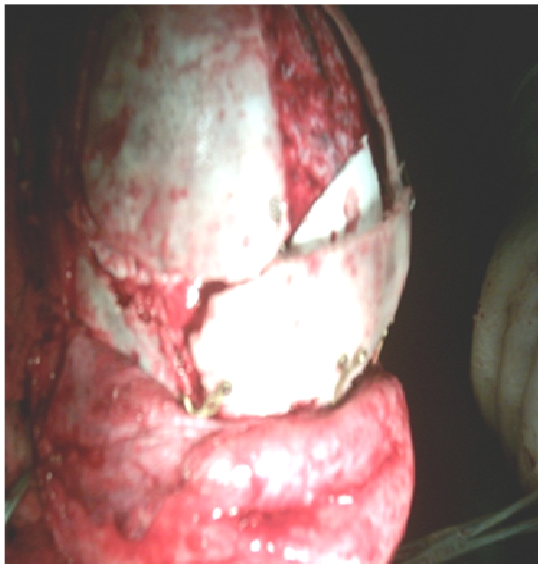


Figure 4



Figure 5

DISCUSSION

The most common complication in patients with symptomatic craniosynostosis is elevated intracranial pressure, which is revealed by diffuse, severe beaten copper pattern in radiograph that originates from the brain pressure on cranium⁽⁹⁾. Astigmatism in uni-coronal synostosis was previously been reported^(10, 11) and there was also one previous report of astigmatism in Apert's, Crouzon's, and a mixture of other craniofacial synostoses⁽¹²⁾. Although visual loss had been documented in these cases and attributed to secondary optic atrophy (due to chronic papilloedema), amblyopia, exposure keratopathy, or a combination of all three^(13, 14), there are no studies to demonstrate the degree of visual loss in craniosynostoses. The optimum time for surgical correction of craniosynostosis is during the first year of life, especially the first

6th month of age; but in this case the rationale for surgery was to improve the visual deficit that was progressively increasing and to prevent the loss of sight.

CONCLUSION

Insidious craniosynostosis is an unusual cause of chronic papilledema in childhood. The papilledema may be resolved and visual loss can be prevented by surgery; the optimum time for surgical correction of craniosynostosis is during first year of life, especially the first 6th month of age.

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