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Management of craniosynostosis at an advanced age: Clinical findings and interdisciplinary treatment in a 17 year-old with pan-suture synostosis

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ABSTRACT

Craniosynostosis is the premature fusion of cranial sutures, occurring at a rate of approximately 1 in 2000 live births; it is usually diagnosed and treated within the first year-of-life. Some diagnoses are delayed and only detected later in childhood or adolescence when symptoms of increased intracranial pressure (ICP) arise such as headaches and vision changes. We present a case of occult craniosynostosis in which a relatively normocephalic 17-year-old male presented with debilitating headaches, optic nerve edema, and developmental delay consistent with probable ICP elevation. CT scan demonstrated pan-suture craniosynostosis. Invasive monitoring confirmed increased ICP for which he underwent cranial remodeling and expansion. While the functional benefits of cranial remodeling are still vigorously debated, this patient's headaches resolved postoperatively. Clinicians should be cognizant of cases of occult craniosynostosis, obtain the appropriate preoperative evaluations, and recognize the utility of cranial remodeling in appropriately selected patients.

Introduction

Craniosynostosis is the premature fusion of one or more cranial sutures. It occurs at an overall rate of 1 in 2000 births [1]. Head shape anomalies or a syndromic diagnosis usually alerts the parent or pediatrician early in infancy to the potential underlying bony pathology. Single suture synostoses result in characteristic head shapes, which facilitate clinical diagnosis. Surgical correction is typically performed within the first year-of-life to expand and normalize cranial shape to accommodate the rapidly growing infant brain [2]. One central controversy in craniofacial surgery is whether cranial remodeling is an aesthetic or functional procedure. Evidence supports opinions on both sides of this contentious debate [3,4]. Cases of untreated craniosynostosis provide some insight into the natural history of this condition [5]. We present a case of pan-suture craniosynostosis with relative normocephaly, which eluded timely diagnosis and treatment. These rare cases serve as a reminder that craniosynostosis can be associated with severe refractory headaches and visual changes in the absence of significant head shape anomalies.

Case report

A 17-year-old male presented with recurrent debilitating headaches and ear and mastoid pain, leading to frequent school absence, grade repetition, and eventual expulsion. As an adolescent he was diagnosed with a Chiari malformation, but the finding of pansuture synostosis remained elusive. He underwent Chiari decompression via a suboccipital craniectomy with a C1 laminectomy. The dura was opened in a Y-shaped fashion. Tonsil tips were found to be ischemic and were elevated with coagulation. Duraplasty using an alloderm patch was performed. The patient was 12-years-of-age at the time of Chiari decompression and did not experience improvement in his headaches postoperatively. He was then evaluated by a second neurosurgeon and was diagnosed with occipital neuralgia and referred for medical management of his headaches. Oral medications failed to improve his symptoms, but illicit marijuana provided some relief. His early medical history was unremarkable; the product of a full-term vaginal delivery, his family reported a normal head shape at birth. He first began to report headaches at 10-years-of-age. Family history was notable for migraines, particularly in his maternal grandmother, but no history of craniofacial anomalies.

At presentation his head circumference was 55 cm (<50th percentile). He had subtle contour abnormalities and asymmetry of the head, with some flattening of the left frontoparietal region, mild

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bilateral mastoid bulging, and a prominence of the right frontal bone. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed pan-suture synostosis and a bony spur at the site of his prior Chiari decompression (Fig. 1). The bony spur was not considered clinically significant as MRI demonstrated adequate decompression with establishment of the cisterna magna. Ophthalmologic evaluation revealed blurring and elevation of the inferior optic disc margins in both eyes with absence of spontaneous venous pulsations, concerning for bilateral optic nerve edema secondary to elevated intracranial pressure (ICP) (Fig. 2A). Optical coherence tomography (OCT) was also performed to determine the thickness of the retinal nerve fiber layer (RNFL), which can aid in assessing the degree of optic nerve edema. OCT revealed a slight blurring of the inferior margin of the optic disc in our patient, consistent with mild optic nerve edema.

Given the controversial indications for delayed cranial remodeling and the potential for confounding causes of his headaches, formal intraparenchymal pressure monitoring was performed using the Codman © Microsensor for 48 h. This revealed elevated intracranial pressures in excess of 20 mm Hg, A wave activity and rhythmic B waves consistent with elevated ICP (Fig. 2B). These findings coupled with the refractory headaches and abnormal ophthalmological exam provided the indication for cranial vault expansion.

The patient underwent craniectomy and posterior cranial vault expansion. Intraoperatively, he was found to have a “tight” brain with immediate expansion and relaxation of the brain upon craniotomy. Preoperative and intraoperative findings were consistent with pan-suture synostosis (Fig. 3A). Cranial reconstruction was performed using stair-step osteotomies to both significantly expand the cranium as well as to maintain cortical coverage (Fig. 3B and C) [6]. The patient spent one night in the ICU and recovered well without any postsurgical complications. He was discharged on postoperative day four. Postoperative CT demonstrated an 18% increase in intracranial volume with complete expansion of the brain to fill the new space (Fig. 4). The change in intracranial volume reflects the bony changes, and was calculated as the difference in volumes contained by the cranium on the pre and post op CT scans.

At his four-week follow-up his headaches had abated, though he still had intermittent mastoid pain. Follow-up ophthalmology evaluation four weeks after surgery demonstrated no change in the subtle optic nerve edema detected preoperatively. At six months follow-up he remained free from headaches, but noted occasional discomfort in his ears and mastoid areas bilaterally.

Discussion

Cases of occult craniosynostosis with relative normocephaly have been previously reported. Martinez-Lage and colleagues documented two such cases in which the diagnosis of craniosynostosis was missed at birth due to negligible skull deformity [7]. Both patients presented years later with elevated ICP and underwent cranial expansion, which alleviated their symptoms.

Our case resembles these accounts of delayed presentation, though this child was older at presentation and had previously undergone Chiari decompression for his symptoms. Our patient had a relatively normal head shape at birth, with only subtle changes apparent in adolescence. The timing of his suture fusion is unclear, though retrospective review of his CT scans at 11-years-of-age demonstrates the synostoses.

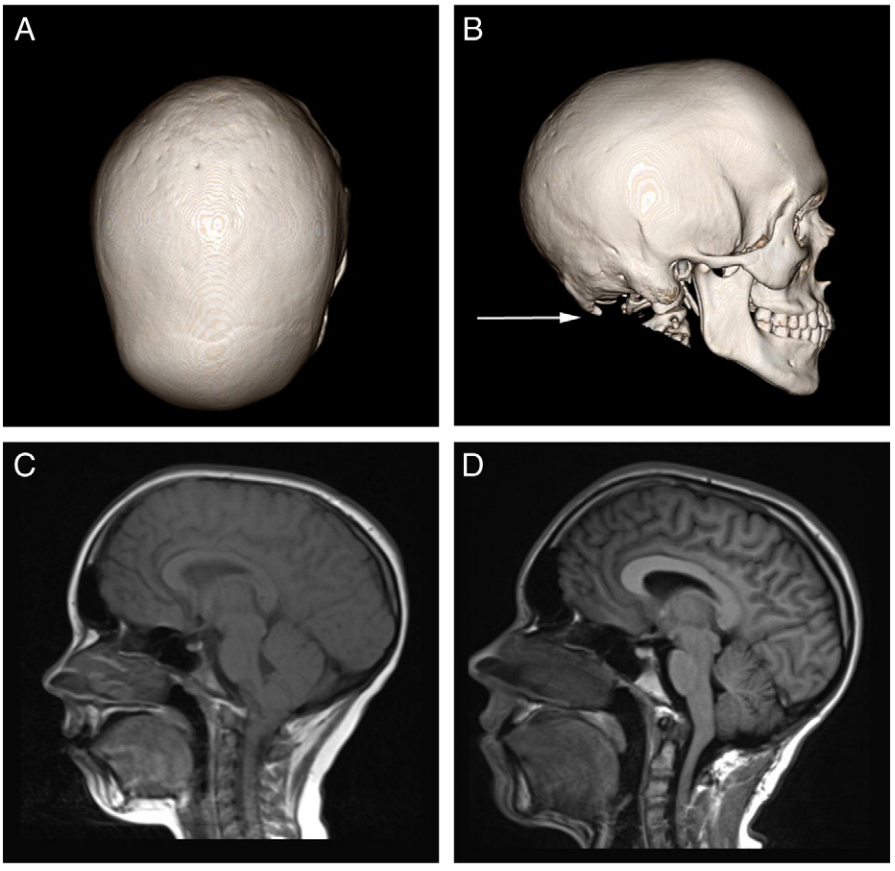

Fig. 1. Pre-operative superior (A) and lateral (B) views of the calvarium on CT scan demonstrating suture fusion and a bony spur at the site of a prior Chiari decompression. T1-weighted MRI scan (C) revealing a Chiari malformation five years prior to his craniofacial workup and post-operative MRI (D) demonstrating decompression of the Chiari malformation.