Pancraniosynostosis following endoscope-assisted strip craniectomy and helmet orthosis for sagittal suture craniosynostosis in a nonsyndromic patient

Case report

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A variety of surgical strategies are used to correct sagittal craniosynostosis. There is increasing experience with the use of endoscope-assisted techniques, although there is no consensus regarding the optimal technique. Pancraniosynostosis occurring after the surgical repair of single-suture craniosynostosis is an unusual complication. The authors describe the case of a nonsyndromic patient who underwent an endoscope-assisted strip craniectomy with the subsequent use of a helmet orthosis for correction of an isolated sagittal suture craniosynostosis. The patient’s early postoperative course was uneventful. Pancraniosynostosis subsequently developed, requiring much more extensive surgical correction. Awareness of this potential complication is necessary to ensure its recognition and appropriate management.

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Key Words • craniosynostosis • pancraniosynostosis • congenital • endoscope-assisted strip craniectomy • helmet orthosis

Craniosynostosis surgery has evolved considerably since first described by Lannelongue and Lane in the late 19th century. Ultimately, the surgical goals remain unchanged: cranial vault expansion to accommodate the growing brain and improved cosmesis. Endoscope-assisted strip craniectomy has been developed in an effort to reduce the need for intraoperative blood transfusions and the duration of hospital stays.

Pancraniosynostosis has been defined as the fusion of 3 or more sutures. It is an uncommon occurrence following the surgical correction of single-suture craniosynostosis, and risk factors for such a complication are unknown. We describe such a case in which a multiple-suture craniosynostosis occurred after an endoscope-assisted craniosynostosis followed by the use of a molding helmet orthosis.

Case Report

History and Examination. A previously well 3-month-old girl with no family history of craniosynostosis presented with scaphocephaly, and an isolated, single-suture sagittal craniosynostosis was diagnosed (Figs. 1 and 2). On examination there was prominent ridging of the sagittal suture and closure of the anterior and posterior fontanels. There was frontal bossing, and her cranial index was 0.70.

First Operation. At 3 months of age, she underwent a successful, uncomplicated endoscope-assisted 3-cm strip craniectomy of the sagittal suture without barrel stave osteotomies. During the operation, the brain did not appear tight.

First Postoperative Course. Postoperatively, she was fitted with a molding helmet orthosis, which she wore continuously except for motor vehicle trips. At the 1- and 3-month follow-up appointments, her cranial index had improved to 0.76 and 0.77, respectively. The skull defects were gradually filling in. After 3 months, it was suggested that she wear the helmet during the night only and that no new helmet should be prepared once she outgrew the current orthosis.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
Six months following the initial surgery, the patient returned to the clinic with a significant, pulsatile bony prominence adjacent to her anterior fontanel. She also had flattening of the left forehead. Her cranial index had increased to 0.8. The overall appearance was that of a left anterior plagiocephaly. On CT scanning (Fig. 3), near-complete fusion of the coronal, sagittal, metopic, and lambdoid sutures was seen.

Second Operation. At the age of 12 months, she underwent a left frontal orbital advancement and cranial vault remodeling with barrel stave osteotomies. A blood transfusion was required intraoperatively; otherwise, the procedure was uncomplicated.

Second Postoperative Course. She was discharged home on the 4th postoperative day. Nine months after the second surgery, her head shape was normal. The forehead contour was excellent. Milestones for speech, comprehension, and gross and fine motor controls were normal for her age. Genetic testing for Saethre-Chotzen syndrome (TWIST gene sequencing and P250R and FGFR3 mutation testing) and Crouzon syndrome (FGFR2 sequencing) revealed no predisposition to craniosynostosis. The pediatric medical geneticist who saw the patient in consultation did not believe that she displayed the clinical characteristics for Pfeiffer syndrome, and thus genetic testing for this condition was not requested.

Discussion

Endoscope-assisted craniectomy followed by the use of a helmet orthosis has been reported as a safe option for correcting craniosynostosis. It is suggested that this strategy is associated with a reduction in blood loss and transfusions, duration of hospital stay, and cost, as compared with other calvarial remodeling procedures. These 6 cited studies include more than 350 patients. Of these patients, 5 had nonsyndromic fusion of new sutures following the initial surgery. Only 1 patient clearly demonstrated pancraniosynostosis.

Reddy et al.12 found 7 nonsyndromic patients with progression from single-suture craniosynostosis to pancraniosynostosis in over 50 years of clinical data. The reported incidence was 1.6%. The most common site for the initially involved suture was sagittal, followed by coronal and lambdoid, which is consistent with the distribution of sutures involved in craniosynostosis. Hudgins et al.5 found 2 nonsyndromic patients among a series of 210 with progressive craniosynostosis, for an incidence of approximately 1.0%. Greene4 documented 3 nonsyndromic children who progressed to pancraniosynostosis following adequate correction of a single-suture synostosis. Beyond these reports, data are scarce on progressive craniosynostosis in nonsyndromic children following successful surgery. Our case adds to this limited list. The studies listed above would suggest that the incidence of pancraniosynostosis appears to be similar in patients treated surgically with or without a postoperative helmet orthosis.

Increased intracranial pressure and its potential sequelae are of potential concern in patients with pancraniosynostosis. In our patient, attempts to document intracranial pressure were not considered since surgery was recommended given the unsatisfactory cosmetic outcome as a result of the pancraniosynostosis.

Conclusions

Progressive craniosynostosis following correction of single-suture craniosynostosis is a rare phenomenon. Our case adds to the limited available literature, particularly cases utilizing an endoscope-assisted technique followed by the use of a helmet orthosis. We recommend follow-up.
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of all patients with craniosynostosis for signs of progressive synostosis.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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