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CASE REPORT

Pediatric/Craniofacial

Unilateral Pterional Polycraniosynostosis Treated with Craniectomy and Helmet Therapy

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Summary: Craniosynostosis is a condition in which one or more of the cranial sutures have fused prematurely, affecting the growth pattern and contours of the infant skull. The pterion is the junction of temporal, frontal, parietal, and sphenoid bones of the skull. We present a case of unilateral pterional craniosynostosis, which was treated with strip craniectomy and helmet therapy. *(Plast Reconstr Surg Glob Open 2017;5:e1245; doi: 10.1097/GOX.0000000000001245; Published online 3 February 2017.)*

Craniosynostosis results from the premature fusion of cranial sutures and affects approximately 1 in 2500 infants.[1] Although the clinical course and treatment of synostosis of the major calvarial sutures (coronal, sagittal, metopic, and lambdoid) are well characterized, there is a clear sparsity of reports describing rarer forms of craniosynostosis and their phenotype.[1–4] We present a case of unilateral pterional polycraniosynostosis. To our knowledge, this is the first report of this pathological entity.

CASE REPORT

A full-term female infant was noted to have a depression of the left temporoparietal skull vault at birth. She was born to a 39-year-old G4P3 mother. Parents were nonconsanguineous and reported no personal or family history of craniosynostosis. Antenatal ultrasound at 19 weeks revealed polycystic kidneys. The child was delivered vaginally without the use of assisted devices and weighed 2560 g. Newborn examination revealed a head circumference of 31.6 cm consistent with microcephaly. A systolic murmur was also detected at birth, which led to the diagnosis of a ventricular septal defect. Genetic testing did not reveal any abnormalities.

The infant was referred to our unit for review at 4 months of age. Physical examination revealed a left temporoparietal skull depression causing marked asymmetry in head shape, a low-set left ear, and a large posterior fontanelle. Fundoscopy was normal. Plain skull x-rays and magnetic resonance imaging failed to demonstrate craniosynostosis. Three-dimensional computerized tomography (3DCT) scan demonstrated synostosis involving the coronal, frontosphenoidal, sphenoparietal, and temporoparietal sutures of the left pterion, and also an anomalous horizontal intraparietal suture synostosis (Fig. 1). The anterior fontanelle, right coronal suture, and lambdoid sutures were patent. The metopic suture was fused, but there was no trigonocephaly to suggest premature fusion.

The child underwent a craniectomy of the fused sutures at 6 months of age via a left hemicoronal incision. A custom-made helmet was fitted by an orthotist on day 4 postsurgery and readjusted on day 9 after resolution of subgaleal swelling. Improvement in head shape was evident at 6 weeks' follow-up. The patient was reviewed regularly in the multidisciplinary craniofacial clinic, and regular 3-dimensional (3D) photographs were taken to visually document progress (Figs. 2, 3). The helmet was worn until the patient was 14 months old. A 3DCT scan performed at the completion of helmet therapy demonstrated improvement in skull contour and only small calvarial defects (Fig. 4) (see figures, **Supplemental Digital Content 1–4**, which show pre- and postoperative clinical photographs and CT scans, ***http://links.lww.com/PRSGO/A379***). In addition, her parents were satisfied with her aesthetic appearance and she is developing normally. Although early results have been satisfactory, ongoing follow-

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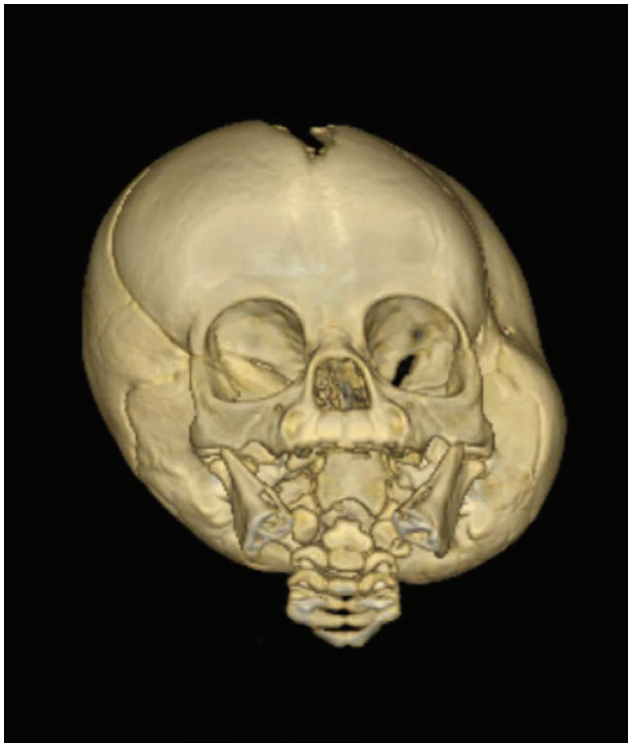

Fig. 1. Preoperative CT scans; child aged 5 months old.

Fig. 2. Preoperative photographs taken at the initial consultation in the craniofacial clinic; child aged 4 months old.

Fig. 3. Photographs taken at 9 months postsurgery child aged 15 months old.

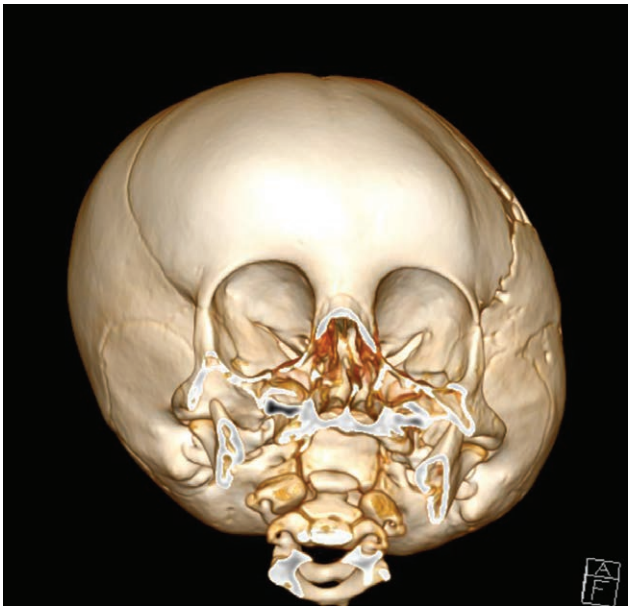

Fig. 4. Postoperative CT scans, performed 9 months postsurgery; child aged 15 months old.

up is necessary to monitor the residual calvarial defects and to determine the long-term outcome of craniectomy and helmet therapy for pterional polycraniosynostosis.

Parental consent was obtained for this case report. Ethics approval was granted by the Sydney Children's Hospitals Network Ethics Committee (CCR2016/13).

DISCUSSION

The pterion is an H-shaped area on the lateral skull formed by the junction of the frontal, temporal, parietal, and sphenoid bones. There are variations in the articulation of these bones with one another, and sometimes a small epipteric bone may be present.[5] Craniosynostosis affecting sutures of the pterion is rare, with only a handful of reports in the literature describing craniosynostosis of the frontosphenoidal,[2] squamotemporal,[3] sphenosquamosal, and sphenoparietal sutures.[1,4] The present case is unique to report craniosynostosis affecting multiple sutures seen at the point of confluence of the pterion, a combination that has not been previously reported. In our present case and others, CT scan enabled the diagnosis of craniosynostosis in the pterional region, which was otherwise not obvious on plain skull x-rays.[3]

Our case patient had a gross ipsilateral temporoparietal skull depression and an ipsilateral low-set ear, which is congruent with the calvarial dysmorphology of ipsilateral flattening reported for synostosis affecting the lateral minor sutures. Ranger et al.[3] found that ipsilateral temporal narrowing was a feature of squamotemporal craniosynosto-

sis. Marucci et al.[2] found that patients with frontosphenoidal synostosis presented with ipsilateral flattened forehead and maxilla, in addition to contralateral nasal root and endocranial base deviation, as well as bony orbital anomalies. Eley et al.[1] and Smart et al.[4] found that ipsilateral occipital flattening was typical of squamosal synostosis even though the clinical findings were highly variable. They noticed that when squamosal synostosis occurred in conjunction with major suture synostosis, the head shape was predicted by the major suture fusion. They also observed that synostosis caused deformity only when the suture contributed significantly to normal growth, with the ultimate deformity dependent on the involvement of other sutures and the presence of syndromes.[1,4]

Our case patient also had a fused anomalous horizontal intraparietal suture. The presence of accessory parietal sutures is extremely rare and may be formed by the partial or total division of the parietal bone. The cause of this division is not well understood but could be due to failure of fusion of the 2 intraparietal ossification centers.[6] Shapiro[7] observed only 3 skulls with accessory parietal sutures out of 25,000 skull radiographs examined in their series, highlighting the rarity of this event. The anomalous suture most commonly crosses the parietal bone horizontally, as seen in our patient, with vertical and oblique courses being less common.[8] The unilateral occurrence of accessory parietal sutures, even when these sutures remain patent, will cause skull asymmetry as there is increased growth perpendicular to the accessory suture, which leads to unequal skull growth between affected and nonaffected sides of the cranial vault.[9]

The final important feature of the presented case is the use of craniectomy and helmet therapy. This is a technique that releases the fused sutures and relies on the rapid brain growth during early infancy to reshape the head contour. The helmet allows for 3D growth and tailored adjustments to the patient's individual course can be made. Patients under the age of 6 months are ideal surgical candidates, and this method has been employed to correct sagittal, coronal, metopic, and lambdoid craniosynostosis.[10] To the best of our knowledge, we present the first case of pterional polycraniosynostosis to be treated with this technique.

CONCLUSION

Given that a literature search has not yielded any similar cases, it is of significance that we have identified this exceptionally unique case of pterional polycraniosynostosis managed with craniectomy and helmet therapy. We hope this case report will prompt further recognition and reporting of rarer forms of craniosynostosis and their clinical course.

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PATIENT CONSENT

Parents or guardians provided written consent for the use of the patient's image.

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