Frontal plagiocephaly secondary to synostosis of the frontosphenoidal suture

Case report

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Frontal plagiocephaly may arise from either synostotic or deformational forces. Deformational causes of frontal plagiocephaly can be distinguished from synostotic causes by differences seen on physical examination, which can then be confirmed by skull x-ray films and if necessary three-dimensional computerized tomography (CT). Unilateral coronal synostosis is the main synostotic cause of frontal plagiocephaly, although it has also been seen with fusion of the frontozygomatic suture. In several syndromes presenting with bilateral coronal synostosis, fusion of the frontosphenoidal and frontoethmoidal sutures is also present.

The authors report, for perhaps the first time, a case showing synostotic frontal plagiocephaly secondary to fusion of the frontosphenoidal suture alone. Although the phenotypic appearance is superficially similar to that seen in unilateral coronal synostosis, analysis of the cranial base shows markedly different effects: angulation of the anterior cranial base with respect to the posterior cranial base away from the synostotic side and angulation of the posterior cranial base with respect to the midpalatal suture also away from the synostotic side. In unilateral coronal synostosis, both angulations are toward the synostotic side. These effects on the cranial base alter its relationship to the cranial vault and the facial skeleton. Most important, frontal plagiocephaly secondary to fusion of the frontosphenoidal suture should not be overlooked as being deformational. Because this fusion is difficult or impossible to visualize by skull x-ray films, three-dimensional CT must be obtained in cases that are not clearly identified as deformational plagiocephaly by physical examination.

KEY WORDS • plagiocephaly • craniosynostosis • coronal synostosis • cranial base

Frontal plagiocephaly in nearly every case has been described in association with unilateral coronal synostosis. In this condition premature fusion occurs at the coronal suture, resulting in a single frontoparietal bone plate ipsilateral to the fused suture, which has a reduced growth. Other cranial vault sutures continue their bone formation because of pressure stimulation from brain expansion. This compensatory growth results in the plagiocephalic appearance. Frontal plagiocephaly may also be caused by deformational forces occurring in utero or at the time of delivery. In typical unilateral coronal synostosis there is ridging of the prematurely fused coronal suture, flattening of the ipsilateral frontal and parietal bones, bulging of the ipsilateral squamous portion of the temporal bone, and bulging of the contralateral frontal and parietal bones. On skull radiographs one observes in addition to sutural sclerosis the so-called “harlequin abnormality.” This abnormality is noted as an elevation of the greater wing of the sphenoid ipsilateral to the fused suture.

Unilateral coronal synostosis is not the only synostotic cause of a frontal plagiocephalic appearance, but the other causes are very rare. Frontozygomatic suture stenosis, for example, can create a frontal plagiocephaly mimicking that seen in unilateral coronal synostosis. In addition, it has also been noted that unilateral coronal synostosis can be associated with stenosis of another portion of the coronal ring, such as the frontosphenoidal or frontoethmoidal sutures, although neither of these other sutural fusions has been noted alone. Bruneteau and Mulliken state that one can clinically separate synostotic and deformational causes of frontal plagiocephaly by careful physical examination focusing on the position and shape of the supraorbital rims, nasal root, ears, malar eminences, chin point, palpebral fissures, and facial height.

We report on a case showing the frontal plagiocephalic appearance that is not secondary to unilateral coronal synostosis but rather is secondary to the fusion of the frontosphenoidal suture alone.
Case Report

This 10-month-old boy was born via a normal spontaneous vaginal delivery; he weighed 3600 g and showed a normal skull shape. At the age of 10 months, however, the mother noted new flattening of the right forehead and orbital rim and exorbitance of the right eye, which led to referral to the craniofacial clinic at the St. Louis Children’s Hospital.

Examination. Examination of the boy revealed frontal plagiocephaly, with the right forehead recessed and the left frontal bone prominent. The cartilaginous nasal tip was deviated toward the right (ipsilateral to the lesion) and the nasal root to the left (contralateral to the lesion). The chin, ear positions, and occiput were symmetrical. The anterior fontanelle was small and midline. He had a head circumference of 45.1 cm (50th percentile), a medial canthal distance of 25 mm (55th percentile), and an interpupillary distance of 45 mm (50th percentile). His gaze continued to be conjugate, but he had significant right eye exorbitance.

Radiological Evaluation. Radiological evaluation included skull films and three-dimensional computerized tomography (CT) with a spiral scanner. The skull films showed no evidence of coronal synostosis despite the typical appearance of frontal plagiocephaly (Fig. 1). Exocranial lateral views (Fig. 2) showed normal sutural pattern of the left coronal, frontozygomatic, sphenotemporal, sphenoparietal, frontosphenoidal, and frontoethmoidal sutures, and likewise showed a normal sutural pattern on the right except for fusion of the right frontosphenoidal suture. The superior view of the cranial vault disclosed the skull shape typical of frontal plagiocephaly (Fig. 3 left), with the right coronal suture open despite the associated plagiocephaly. In contrast to unilateral coronal synostosis, the anterior fontanelle in our patient is midline rather than shifted toward the fused side. Figure 3 right shows the frontal view of the skull, revealing the harlequin abnormality secondary to elevation of the greater sphenoid wing.

Fig. 1. Skull radiographs showing slight elevation of the sphenoid wing without a definite harlequin deformity (left) and open coronal sutures (right).

Fig. 2. Three-dimensional computerized tomography showing exocranial views of the calvaria. Left (left) and right (right) lateral views show normal sutures on the left, but fusion of the frontosphenoidal suture on the right.

Fig. 3. Skull radiographs. Left: Superior view showing the cranial vault. This view illustrates the flattening of the frontal and parietal bones on the ipsilateral side, a nearly closed anterior fontanelle, and compensatory bulging of the contralateral frontal bone. Note that the right coronal suture is open despite the associated plagiocephaly. The anterior fontanelle is midline rather than shifted toward the synostosed suture as seen in unilateral coronal synostosis. Right: Frontal view showing the harlequin abnormality on the right side, which is secondary to elevation of the sphenoid wing.

Fig. 4. Three-dimensional computerized tomography scans. Left: Endocranial base view showing that the angulation of the anterior cranial base with respect to the posterior cranial base is 9° away from the synostotic side. In unilateral coronal synostosis, the angulation is 11° toward the synostotic side. Right: Exocranial base view showing that the angulation between the midpalatal suture and the posterior cranial base is 8° away from the synostotic side. In unilateral coronal synostosis, the angulation is 7° toward the synostotic side.
wing. Further analysis of this deformity requires evaluation of the skull base (Fig. 4 left). In this patient, the angulation of the anterior cranial base with respect to the posterior cranial base is 9° away from the side of the synostosis (contralateral). This is different from that noted in unilateral coronal synostosis, in which the same angle is 11° toward the synostotic side (ipsilateral). In a similar fashion, the angle between the midpalatal suture and the posterior cranial base on the exocranial base view (Fig. 4 right) in this patient is 8° away from the synostotic side. In unilateral coronal synostosis, the same angle on exocranial base views is 7° toward the synostotic side.

Operative Findings. The patient was placed supine with the head slightly flexed. A bicoronal incision and a supraperiosteal dissection were then performed, producing a large bifrontal flap. The high resolution three-dimensional CT findings were confirmed intraoperatively (Fig. 5). The right frontosphenoidal suture was closed, but the right sphenotemporal, sphenoparietal, coronal, frontoethmoidal, and frontozygomatic sutures all were open. The surgical correction of frontal plagiocephaly was performed as previously described.9

Postoperative Treatment. Postoperatively, a follow-up three-dimensional CT scan was obtained. Histological analysis of both coronal sutures and the right sphenoparietal, sphenomental, and frontozygomatic sutures was normal. The right frontosphenoidal suture was replaced by bone from intramembranous ossification consistent with early synostosis.

Discussion

This case report is the first published account of a patient with the appearance of frontal plagiocephaly secondary to fusion of the frontosphenoidal suture alone. The resulting frontal plagiocephaly is similar to that noted for unilateral coronal synostosis; however, such a sutural fusion did not exist. Frontal plagiocephaly secondary to unilateral coronal synostosis and frontosphenoidal synostosis both cause an impairment of the ventral expansion of the anterior cranial fossa, which results in its compression.9 This anterior cranial base deformity results in a recessed brow and a flattened forehead. The growth vectors of the frontal lobe become redirected superiorly, inferiorly, and contralaterally. The superior pressure causes the elongation of the forehead, whereas the inferior pressure creates a deformity of the subjacent middle cranial fossa, resulting in ventral bowing of the greater wing of the sphenoid. Exorbitance in both conditions is caused by recession of the superior orbital rims, shortening of the lateral wall of the orbit, and direct encroachment upon the orbital volume by the middle cranial fossa contents. Other abnormalities that are seen in frontal plagiocephaly from both causes include orbital dystopia and occasional midface asymmetries probably secondary to torque in the clivus induced by the synostosis.1, 8–10,13–15,17,18

One phenotypic difference noted is in the direction of deviation of the nasal root. Whereas in this patient the nasal root deviated away from the involved side, in unilateral coronal synostosis the deviation is toward the involved side.16 To understand this change in the direction of the projection of the nasal root and the slight changes in facial features, it is best to reevaluate the three-dimensional CT. In evaluating the endocranial base view, this patient with frontosphenoidal synostosis shows angulation of the anterior cranial base away from the synostosis, whereas in unilateral coronal synostosis, the angulation is toward the synostosis. This angulation of the cranial base causes the nasal tip to project in the opposite direction.

The etiology of the plagiocephaly noted in this case is unclear. Although the exact etiology of the suture fusion in unilateral coronal synostosis is not known, Persing and Jane,11,19–21 among others,3,7 have suggested a fairly reasonable explanation of the bone plate growth pattern and the resulting skull shape. However, the marked similarities and slight differences between the phenotype of unilateral coronal synostosis and frontosphenoidal synostosis may only be superficial. Clearly the two causes of frontal plagiocephaly have effects on the cranial base in entire-
ly different directions, with unilateral coronal synostosis causing angulation of the anterior cranial base toward the fused suture and frontosphenoidal synostosis causing angulation away from the fused suture. A similar pattern is seen in terms of the angulation of the posterior cranial base relative to the midpalatal suture in the two synostoses. The etiology of this entirely different effect is unclear, and therefore we believe this is the first reported case of such a finding.

Examples of two entirely different disorders producing superficially similar cranial anomalies are Crouzon’s and Apert’s syndromes. It has been known that the cranial anomalies in these two syndromes are similar, and a hypothesis of fusion of the entire coronal ring (bilateral coronal, frontosphenoidal, and frontoethmoidal sutures) had been generally accepted as the etiology of the turbrachycephaly noted in both conditions. More recent evidence now shows that this phenotypic similarity results from very different cranial processes, with Apert’s syndrome initially showing only primary closure of the coronal sutures and a midline calvarial defect, whereas Crouzon’s syndrome initially shows multiple premature fusions of sutures and synchondroses. After the 1st year of age, however, the two both show similar closure of sutures and fontanelles, although they have drastically different effects on the cranial base: clival kyphosis in Crouzon’s syndrome versus platybasia in Apert’s syndrome.

Knowledge of the differing effects on the cranial base in the case of unilateral coronal synostosis versus frontosphenoidal synostosis as well as in the example of Apert’s and Crouzon’s syndromes is important from a treatment viewpoint, because the facial skeleton and cranial base cannot be expected to normalize completely after surgery, and further corrective surgery may be needed to deal with these associated abnormalities.

Because plain skull x-ray films may not reveal synostosis of the frontosphenoidal suture, a misdiagnosis of the plagiocephaly as deformational frontal plagiocephaly can occur. Indeed, the three-dimensional spiral CT was diagnostic of the frontosphenoidal suture involvement, whereas the plain x-ray films revealed no abnormality. This disparity emphasizes the need to obtain such a study as part of the preoperative evaluation. The need to distinguish the various synostotic causes of frontal plagiocephaly from deformational frontal plagiocephaly is critical because operative intervention is required in synostotic plagiocephaly to avoid the potentially serious complications of progressive frontal plagiocephaly, whereas deformational plagiocephaly usually improves without operative intervention.

References

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