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TRIGONOCEPHALY IN DIZYGOTIC TWINS - A CASE REPORT

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Trigonocephaly denotes the calvarial deformity caused by premature closure of the metopic suture (metopic synostosis). The frequency of craniosynostosis in general is estimated to be 0.4 per 1000 live births, and trigonocephaly accounts for 5% of all craniostenoses, meaning that this condition is fairly rare. Several studies disclosed that craniosynostosis in twins is a very rare occurrence among craniofacial anomalies. We present a rare case of trigonocephaly in twins where surgery yielded cosmetically satisfactory results.

Key words: Trigonocephaly ▪ Twins ▪ Surgical correction

Introduction

The metopic suture is unique among the calvarial sutures in that it is the only suture that completely disappears and is unidentifiable in the mature adult skull. Evidence suggests that metopic suture closure is complete by 6-9 months of age in 70-100% of children (1, 2). The remainder of calvarial sutures begin closing between ages 26 and 39 years and remain identifiable throughout life. Metopic synostosis denotes the premature closure of the metopic suture. It should be differentiated from trigonocephaly, a term first coined by Welcker in 1862 to decribe an observed calvarial malformation. The incidence of trigonocephaly has been estimated to be between 1 in 2500 and 1 in 15000 births and has been reported to comprise 10 to 20% of patients referred to craniofacial centers. The observation that some infants, identified as having in-utero restraint, are subsequently born with metopic synostosis has led to the hypothesis that this condition may commonly result from fetal constraint. In addition,

conditions that are associated with decreased brain development and microcephaly, such as trisomy 13, can also be associated with early metopic sutural closure. Trigonocephaly has also been noted in twins, both with and without concordance (3). We report of a rare case of trigonocephaly in dizygotic twins.

Methods

Six month-old dizygotic twin boys were admitted for corrective procedure for prominent trigonocephaly. Figure 1 reveals the disfiguring frontal keel in twin infants with trigonocephaly.

Figure 1 *Preoperative appearance of the 3 month-old boys*

Lateral- oblique view accents the deformity (Figure 2).

Figure 2 *Lateral oblique view demonstrating the severe frontal bulging.*

Figures 3 Depicts a preoperative CT scan with 3D SSD reconstruction.

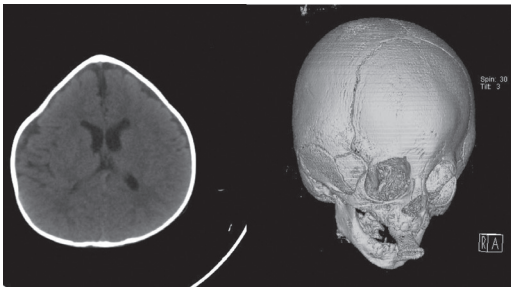

Figure 3 *Non-enhanced CT scan and a SSD reconstruction*

Both boys were subjected to corrective procedure with both frontal and supraorbital remodeling. A bicoronary skin incision was made and a bicoronary skin flap was developed extending to below the supraorbital rims. A bicoronary craniotomy extending beyond the frontiers of coronal suture was fashioned and the frontal bone flap was remodeled. In the subsequent phase, supraorbital rim osteotomy was carried out. Fronto-orbital advancement and cranial reshaping were performed and the bones were stabilized with titanium miniplates. Next, release and forward rotation of the temporal muscle was performed. Figure 4 depicts some of the operative steps performed.

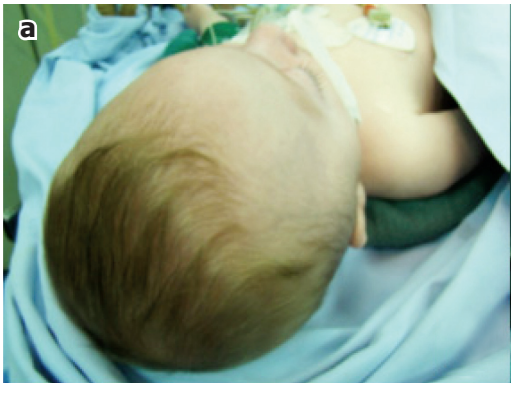

Figure 4 *Intraoperative steps during a trigonocephaly repair: a) patient as positioned before draping, b) a bi-frontal skin flap was developed and rolled downwards exposing a frontal narrowing c) the bifrontal bone flap is elevated d) developing a plane around the orbital arcs, preparing for supraorbital osteotomy e) bone fragments before remodeling f-h) Fronto-orbital advancement and cranial reshaping were performed and the bones were stabilized with titanium miniplates and sutures*

Results

The aforementioned procedure resulted in a cosmetically satisfactory outcome. Figures 5 and 6 reveal the postoperative appearance of these patients.

Figure 5 *Early postoperative appearance*

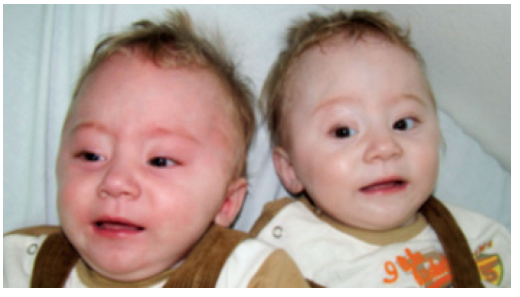

Figure 6 *Appearance 5 months after the surgery*

Figure 7 (a and b) show the postoperative CT appearance (10 months after the surgery).

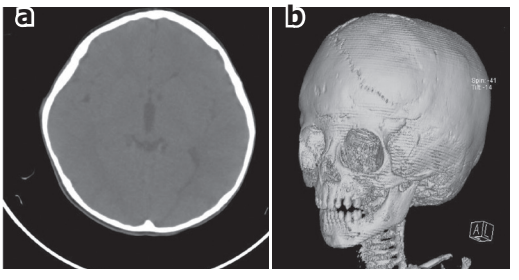

Figure 7 *Non-enhanced postoperative CT scan (a) and Postoperative SSD 3D reconstructed CT (b)*

Discussion

According to Hunter and Rudd, the frequency of craniosynostosis is estimated to be 0.4 per 1000 live births. Trigonocephaly accounts for about 5% of all craniosynostoses reported in the literature (4). Di Rocco reported an increasing frequency of trigonocephaly (420% increase over a 20 year period as compared to other craniosynostoses) (5). Craniofacial anomalies occur with increased frequency in twins as compared with single births. Keusch et al. (6) reported on 35 twin pairs in a series of 1114 congenital craniofacial deformity patients (which accounts for 4%) including 3 craniosynostosis patients. From their survey it appeared that craniosynostosis in twins is a very rare occurrence among craniofacial anomalies. On the contrary, several previous studies have dealt with craniosynostosis in twins and they have presented frequencies ranging from 2.4 to 19.4 percent. Among these studies oxycephaly was reported more frequently. Lajeunie et al. analyzed a series of 1,713 patients with craniosynostosis, hospitalized between 1976 and 1996. They identified 237 patients with metopic synostosis with the male-to-female ratio being 3.3:1. There was no maternal or paternal age effect. The frequency of twinning was 7.8% with two concordances for metopic synostosis in two monozygotic twin pairs. The series was divided into two groups: nonsyndromal trigonocephaly (n = 184) and trigonocephaly associated with other malformations (n = 53). The second group included 13 cases of well-delineated syndromes and 40 cases of trigonocephaly associated with one or more malformations, but without any known syndrome, that could be undelineated syndromes. These groups differed significantly in their mental prognosis (7). Generally speaking, indications for surgery in craniosynostosis patients include cosmetic considerations and prevention of neurological injury (8, 9). Although it was previously generally believed that patients with typical trigonocephaly rarely exhibited clinical symptoms (10), recent papers (11,

12) have reported that such patients experience developmental delays. We presented a rare case of trigonocephaly in twins where surgery yielded cosmetically satisfactory results.

Conflict of Interest: The authors declare that they have no conflict of interest. This study was not sponsored by any external organization.

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