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Metopic and Sagittal Suture Cranioschisis Mimicking Bilateral Parietal Bone Agenesis in a Hydrocephalic Newborn: A Case Report

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Abstract

Introduction: Absence of cranial bones is a rare condition that is incompatible with life. Parietal bone agenesis has rarely been reported in the literature. We performed a search for bilateral parietal bone agenesis associated with hydrocephalus and found that this was one of the few cases in the literature.

Case Report: We presented a case with metopic and sagitta suture cranioschisis mimicking bilateral parietal bone agenesis (University of Izmir of Medicine, Turkey, 05.01.2016). There is a large defective area in our case similar to parietal bone agenesis, however, it has not anencephaly. Due to all of these aspects, our case is secondary cranioschisis. We have planned three stages of treatment for this case: 1) treatment of hydrocephalus, 2) tension bands to close the cranial bones and 3) cranioplasty.

Conclusions: In the determination via 3D cranial tomography the presence of the parietal bones is important because, in case of agenesis the tension band does not benefit and on the contrary it creates extra stress on cerebral tissue.

Keywords: Cranioschisis, Metopic, Parietal Bone, Sagittal, Suture

1. Introduction

Absence of cranial bones is a rare condition that is incompatible with life. Parietal bone agenesis has rarely been reported in the literature (1). We performed a search for bilateral parietal bone agenesis associated with hydrocephalus and found that this was the one of few cases in the literature. In these groups of patients, determination via 3D cranial tomography of presence of the parietal bones is important and in case of agenesis we must use an orthosis to approximate the calvarial bones. There are reconstructions of CT scans of the skull, which would reveal the anomaly clearly. There is a large defective area in our case similar to the parietal bone agenesis. However, it has not anencephaly. Due to all of these aspects, our case is secondary cranioschisis. This case is worth presenting because it can create a great defect similar to parietal bone agenesis, beginning secondary of hydrocephalus and treatment approaches.

2. Case Report

In our hospital (University of Izmir of Medicine, Turkey, 05.01.2016), a 27-week-old male infant was born preterm weighting 815 grams and was one of twins who was born by cesarean section. The infant was found with a 12 × 8 cm calvarial defect on his vertex. The values were measured on the 3D tomography apparatus by ruler mode, which was controlled and calibrated by the same physician. The cranial tomography of the patient with significant breathing difficulty at birth revealed germinal matrix hemorrhage in the right lateral ventricle and hydrocephalus. The infant was found to either parietal bone but metopic and sagittal suture cranischisis mimicking parietal bone agenesis revealed on cranial 3D tomography. The thickness of the cerebral parenchyma was 3 mm. on tomography due to hydrocephalus. Cranial tomography in the 2nd week after birth revealed bilateral germinal matrix hemorrhage and a 2 cm left intraparenchymal hematoma. The brain parenchyma was in the form of a thin strip due to hydrocephalus. The baby showed good activity and there was marked protuberance of the front and rear fontanelle. Surfactant treatment was started for breathing difficulty. He was followed up with ventricular taps for the hydrocephalus until he reached 2000 g, taking into account the low weight and the germinal matrix hemorrhage. A ventricular tap was performed every two days and transcranial ultrasound was used as a follow up. A ventriculo-peritoneal shunt was inserted from the right when he reached 2000 g in weight 10 weeks after birth as the ventricular puncture was clear and the Pandy test result was negative. The scalp at the calvarium dome collapsed due to metopic and sagittal suture cranioschisis in the period after the shunt and the cranium had a 'cup' appearance (Figure 1). The tomography shows the widened brain parenchyma, defect due to the metopic

and sagittal suture cranioschisis more clearly (Figure 2A, 2B, 2C, 2D,). Tension was applied with adhesive bands to approximate the two temporal bones and frontal-occipital bone to treat the 'cup-shaped head' that developed due to the metopic and sagittal suture cranioschisis. A follow up continues with these bands.

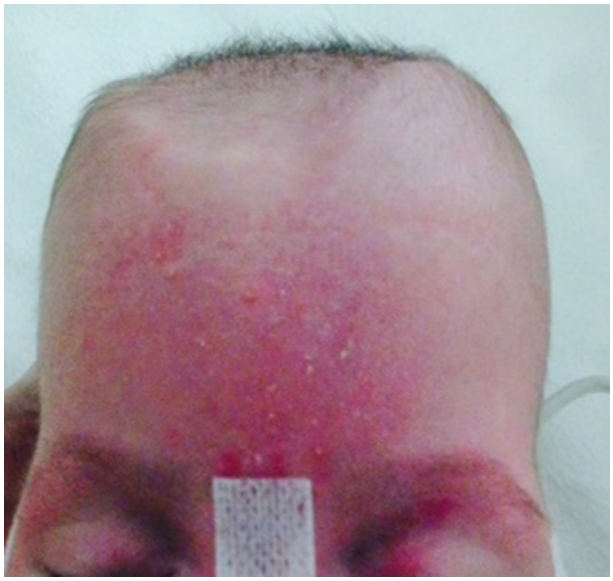

Figure 1. The Cranium Had a 'Cup' Appearance After Ventriculoperitoneal Shunt

Table 1. Case Summary

Age	**27 Week Old**
Weight	815 gr
Delivery mode	Cesarean section
Diagnosis	12 × 8 cm calvarial defect-bilateral parietal bone agenesis-hydrocephalus-germinal matrix hemorrhage
Treatment	V-P shunt-adhesive tension bands to temporal and frontooccpital bones

3. Discussion

Congenital anomalies of calvarial bones are extremely rare. These lesions are divided into three subjects: 1) cranioschisis, 2) craniodysostosis and 3) foramina parietalia permagna. Parietal bone agenesis may be a fourth entity (2).

Absence of the cranial bones is a rare condition that is incompatible with life. Parietal bone agenesis has rarely been reported in the literature (1). However, the more interesting condition of bilateral parietal bone agenesis associated with hydrocephalus has been reported once. That case was also managed conservatively with an orthosis, such as in our case.

Bilateral parietal bone agenesis is rare, however, various anomalies involving the parietal bones have been reported. One of these anomalies is a wide parietal defect reported by Nikolic et al. in an 80-year-old female. The condition is known as Foramina Parietale Permagna. The defect was covered with a fibrous membrane and normal scalp tissue (3). This abnormal parietal bone structure can be a useful tool in forensic cases.

Nicolic et al. reported another parietal bone abnormality in another article. This was an 'interparietal bone' between the two parietal bones, continuing with the frontal region bones. This abnormality was identified during the autopsy of a 54-year-old male who died after head trauma and may again be significant in forensic medicine practice (4).

Fenton et al. reported a case of plagiocephaly due to an extra suture line located in the parietal bone (5). Angonese et al. found two parietal bones, which were both divided into two. They first thought this was a fracture line during a post-traumatic evaluation in a male infant born at 37 weeks. The origin of this rare anatomic variant is not clear (6).

The above are the noteworthy parietal bone anomalies in the literature. Bilateral parietal agenesis has been reported very rarely in the literature. It is most commonly mentioned in a syndrome named 'cleidocranial dysplasia'. This syndrome has been reported a couple of times in the literature. Prominent clinical signs are late closures of the cranial sutures and fontanelles, hypoplastic clavicle and a short stature. This appearance was present in a case of Broeks et al. along with a lack of both parietal bones (7).

Shen et al. reported three children with the 'cleidocranial dysplasia' syndrome. Common characteristics were hypoplastic clavicle, lack of calvarium, open fontanelle and extra teeth. Bone mineral densities, the free thyroxine and TSH levels of these children were found to be normal. A mutation in the RUNX2 transcription factor, a major factor or bone differentiation, is one of the causes of this syndrome. One of the three children had mental retardation. The disease can be transmitted with autosomal dominant inheritance. Two of the cases were hereditary and one was sporadic (8).

The 8th day of gestation is very important regarding occurring cranioschisis. If in the 8th day of gestation, the teratogens affect somits where cephalic localization, the somits becomes necrotic and the anterior part of the neural tube is not closed, which causes the degeneration of

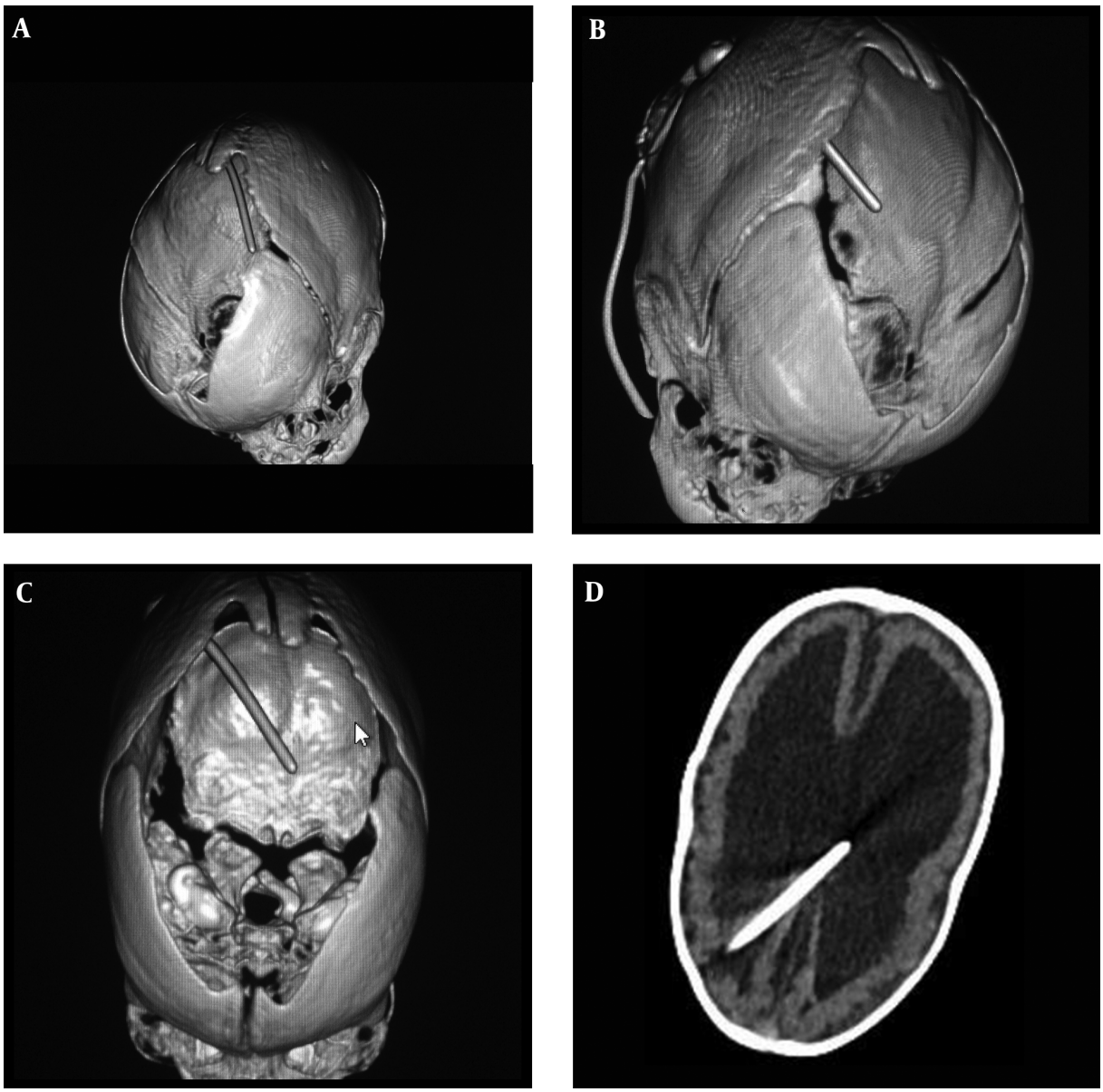

Figure 2. A, the oblique view from the left upper side, 3D cranial tomography; B, the oblique view from the right upper side, 3D cranial tomography; C, the view from the upper side, 3D cranial tomography; D, after ventriculoperitoneal shunt, cranial tomography.

neural tube. Anencephaly is a component of cranioschisis (1).

Our case is metopic and a sagittal suture cranioschisis is to be asscociated with hydrocephalus. Anencephaly is not apperad because cranioschisis occur secondarily and the size of the defective area on the vertex mimicks biparietal bone agenesis. Due to all of these aspects, our case is secondary cranioschisis. We have planned three stages of treatment for this case: 1) Treatment of hydrocephalus, 2) Tension bands to close the cranial bones and 3) Cranioplasty. Determination via 3D cranial tomography of presence of the parietal bones is important because, in case of agenesis, tension bands does not benefit on the contrary it creates extra stress on cerebral tissue.

Cranioplasty is clearly required for the permanent treatment of this disorder however, cranioplasty can only be considered after the age of 6. Ropper et al. used a full layer autologous bilateral parietal bone graft and placed

it on the frontal region during cranioplasty in a 6-year-old patient with a bilateral wide frontal defect. The donor area recovered through fusion of the bone particles taken from the graft's endocortical and ectocortical surface (9).

We used an approximating orthosis for the still flexible calvarial bones in order to facilitate cranioplasty. We applied tension with two adhesive band strips to approximate the temporal bones and two adhesive band strip to approximate the frontal-occipital bones. This system is of course open to development with hard material. The impressive part of our work is that this is very rare case and the treatment modality of tension bands is very rarely used. The data extract's center characteristic is governmental and not referral.

3.1. Conclusion

Our case is metopic and a sagittal suture secondary cranioschisisl which is associated with hydrocephalus. The calvarial shape that developed after the CSF pressure that decreased following the V-P shunt can be named 'cup cranium'. Our treatment approach is to primarily use an orthosis to approximate the calvarial bones and then cranioplasty at the appropriate age. Determination via 3D cranial tomography of presence of the parietal bones is important because, in case of agenesis, tension bands does not benefit, on the contrary it creates extra stress on cerebral tissue. Neurological and systemic pathologies associated with the defect should also be monitored.

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Footnote

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References

1. Moscote-Salazar LR, Castellar-Leones SM, Alcala-Cerra G, Gutierrez-Paternina JJ. Congenital agenesis of cranial bones associated with hydrocephalus: a case report [in Spanish]. *Bol Asoc Med P R.* 2013;**105**(1):54–6. [PubMed: 23767388].
2. de Heer IM, van Nesselrooij BP, Spliet W, Vermeij-Keers C. Parietal bone agenesis and associated multiple congenital anomalies. *J Craniofac Surg.* 2003;**14**(2):192–6. [PubMed: 12621289].
3. Nikolic S, Zivkovic V, Strajina V. Foramina parietalia permagna: case report [in Serbian]. *Srp Arh Celok Lek.* 2012;**140**(9-10):658–61. [PubMed: 23289287].
4. Nikolic S, Zivkovic V, Jukovic F. Interparietal bone in forensic practice: case report [in Serbian]. *Srp Arh Celok Lek.* 2010;**138**(11-12):764–7. [PubMed: 21365891].
5. Fenton RA, Kinsella CR, Cray JJ, Cooper GM, Losee JE, Grunwaldt LJ. Divided parietal bone in plagiocephaly. *J Craniofac Surg.* 2011;**22**(1):355–8. doi: 10.1097/SCS.0b013e3181f7e151. [PubMed: 21239935].
6. Angonese A, Sonnaert M, Rassart A, Gauquier N, Cavatorta E. Skull fracture or bipartite parietal bone [in French]. *Arch Pediatr.* 2010;**17**(4):391–3. doi: 10.1016/j.arcped.2010.01.009. [PubMed: 20202802].
7. Broeks I, Veenstra-Knol IE, Kamps AWA. A rare presentation of cleidocranial dysplasia. *Case Reports.* 2012;**2012**(1):0320126101. doi: 10.1136/bcr-03-2012-6101.
8. Shen Z, Zou CC, Yang RW, Zhao ZY. Cleidocranial dysplasia: report of 3 cases and literature review. *Clin Pediatr (Phila).* 2009;**48**(2):194–8. doi: 10.1177/0009922808323107. [PubMed: 18832541].
9. Ropper AE, Rogers GF, Ridgway EB, Proctor MR. Repair of a large congenital frontal bone defect with autologous exchange cranioplasty. *J Neurosurg Pediatr.* 2010;**6**(5):464–7. doi: 10.3171/2010.8.PEDS10140. [PubMed: 21039170].