Article

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Association of abnormal metopic suture causing hypertelorism, interfrontal encephalocele with craniofacial cosmetic deformity associated with myelomeningocele: management literature review

Willem Guillermo Calderon-Miranda[1], Guru Dutta Satyarthee[2], Huber Padilla-Zambrano[3], YanCarlos Ramos-Villegas[3], Hernando Raphael Alvis-Miranda[4], Joulen Mo-Carrascal[5], Johana Maraby[5], Luis Rafael Moscote-Salazar[6]

[1]Resident of Radiology, Universidad Nacional Autonoma de Mexico, Ciudad de Mexico, MEXICO
[2]Neurosurgeon, Department of neurosurgery, Neurosciences Cengtre, AIIMS New Delhi, INDIA
[3]University of Cartagena, Cartagena de Indias, COLOMBIA
[4]Resident of Neurosurgery, University of Cartagena, Cartagena de Indias, COLOMBIA
[5]Physician, University of Cartagena, Cartagena de Indias, COLOMBIA
[6]Neurosurgeon, RED LATINO, Latin American Trauma & Intensive Neuro-Care Organization, Bogota, COLOMBIA

Abstract: Myelomeningocele may be associated with other neural and extraneural anomalies. Authors present association of metopic suture abnormality, an interfrontal encephalocele with widening of metopic suture and abnormal shape frontal bones in the forehead in those associated with hydrocephalus. Authors describes two neonates with interfrontal encephalocele, representing first series reporting in neonate. Management and pertinent literature is briefly discussed.

Key words: metopic sututre abnormality, hypertelorism, meningomyelocele, hydrocephalus, neonate

Introduction

The meningomyelocele may be associated with Chiari malformation, hydrocephalus, and corpus callosal agenesis. The hydrocephalus may produce large bulging forehead with sunset sign. (1-5) However, abnormal persistence of widely spaced unfused metopic suture is extremely uncommon. The widely opened metopic suture extends superiorly to the anterior fontanelle and inferiorly also extends to the roots of nose causing

hypertelorism and moulding of frontal bones. (4-8) through the metopic suture defect, the brain parenchyma may herniates and herniated brain may remain vulnerable to external injury and associated consequences. The hydrocephalus is postulated to potentially aggravate further herniation of brain. These calvarial and brain parenchymal abnormity cases may not require surgery, and the repair of meningocele with ventriculoperitoneal shunt surgery may be sufficient. Such cases were reported in late childhood. (8) However, both of our current cases were in the neonatal age, representing first of it's in the western literature. Author reports two interesting cases.

Here, we present series of five MMC children with interfrontal encephalocele and unfused wide metopic suture and describe the typical feature of this rare abnormality, the probable differences with other MMC patients, associated problems, and management.

Case illustration

Case #1

Newborn male with evidence of myelomeningocele in sacral region. Born at 32.8 gestation weeks with a weight of 2900 grams and a cephalic perimeter of 53 centimeters, from a 26 years old mother, G6 P3 A3 C3, the six pregnancies with differents fathers, prenatal controls since the first month of pregnancy, attending 8 consultation in all, she performed 4 prenatal ecographies, of which none of them made the diagnosis of myelomeningocele, ambiguous genitals, bilateral varus equine foot and/o metopic suture defect in U form. Serologic evaluation for HIV, syphilis, toxoplasma and HBV were non-reactive.

To physical examination feels a distracted metopic suture, observes ambiguous genitals (scrotum bifidus, hypospadia and micropenis), bilateral varus equine foot, bilateral simian fold and myelomeningocele with fistula, which was operated immediately. Simple skull TC with 3D reconstruction evidences metopic suture defect in U form. (Fig. 1). Cerebral and total spine MRI shows inconspicuous asymmetric cerebellar atrophy predominating on the left side, in the right occipital lobe the rotations are disorganized, there is hypoplasia of the corpus callosum, syringomyelia, low tonsils, supratentorial hydrocephalus and sacral meningocele. Correction is performed through resection of ruptured myelomeningocele, dural graft and fasciocutaneous flap, hemodynamically stable evolution after 16 days of prophylactic scheme for neuroinfection is ordered the discharge.

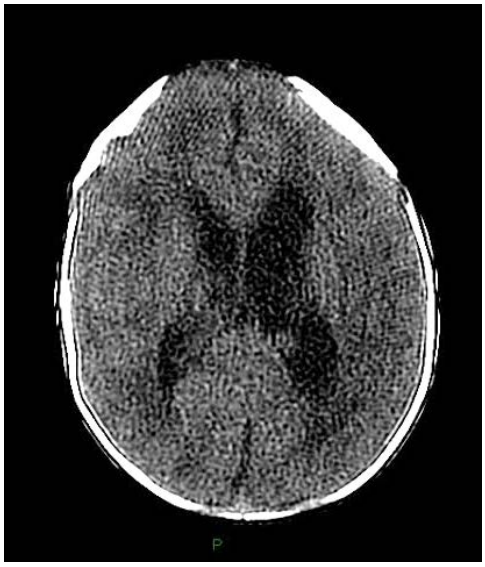


Figure 1 - CT scan showing an abnormal metopic suture

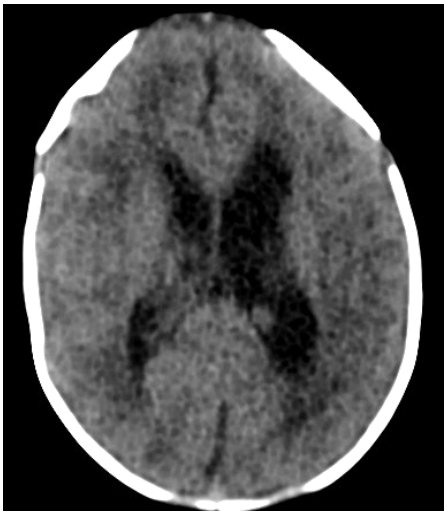
Case # 2

Full term newborn male, product of a fifth pregnancy, from a 41 years old mother, apparently healthy. Pregnancy evolved with adequate prenatal care. Born by Caesarea, punctuation Apgar was 9/10 with appropriate weight to the age, (3850 grams). Physical examination was found to be defective in the lumbar region, suggestive of myelomeningocele. In the cephalic region is evidenced dolichocephalic skull shape, with protrusion on the forehead. Brain CT scan was performed which showed partial agenesis of the frontal bone. (Figure 2 A and B). It neurosurgery performed to correction of neural tube defect. The patient presented communicating hydrocephalus a week later which required development ventriculoperitoneal shunt placement, with satisfactory postoperative evolution.

Discussion

The metopic suture is also called median frontal suture, often associated with frontal sinus hypoplasia or agenesis. Metopic suture extends through the nasion to the bregma, in the midline across the frontal bone and often remain incomplete and usually fuses by around nine months after birth. (1,2)

Rarely, the metopic suture can persist as an anatomical variant, which can be mistaken for fracture of the frontal bone, which is differentiated radiologically the metopic sutures lies in the midline and sutural interdigitations. (3) A premature fusion of the suture is termed metopic synostosis which characterically result into trigonocephaly. (2)



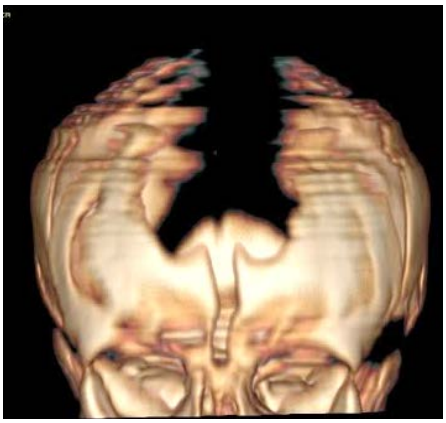


Figure 2 A and B - CT scan showing an abnormal metopic suture

Myelomeningocele develops during the neurulation phase of embryonic life due to failure in posterior neuropore closure. Lacunar skull caused by membranous bone dysplasia typically have well-defined lucent parts and other are frontal dysplasia cranium bifida, [4, 5, 6].

Nejat et al postulated the persistent of lacuna in frontal bones in the region of metopic suture may cause scalloping of the frontal bone with predisposition to persist

widely opened metopic suture, pulsation of herniated neural tissue causes abnormality in the metopic suture region also causing cosmetic deformity. (7)

The metopic suture abnormality progression is usually halted with ventriculoperitoneal shunt surgery and usually separate surgery for metopic suture is not advocated, however, rarely cases with severe hypertelorism and cosmetic deformity may be advised. (7)

Nejat et al. analyzed five children with mean age of six month and all were male. Meningomyelocele was located in the lumbar and lumbosacral areas. Neurological deficits included a spectrum of presentation from sphincter disturbance to paraplegia. All had associated hydrocephalus and required CSF shunt surgery. Cranial neuroimaging showed intracranial, interfrontal encephalocele, asymptomatic Chiari malformation, and corpus callosum agenesis in five, five, and three patients, respectively. These cases had anterior fontanel anomalies called as interfrontal encephalocele, associated with open metopic suture extending widely to the nasal radix producing hypertelorism accompanied by interfrontal herniation of frontal lobes. The associated hydrocephalus exaggerates the interfrontal encephalocele and shunt surgery ameliorates interfrontal encephalocele. However, some patient may have surgical correction of metopic suture for cosmetic (7).

Conclusion

Authors reports two cases of forehead interfrontal encephalocele in two neonate, who had associated myelomeningocele. Patients had widened metopic suture and association with herniation of frontal lobe through the midline calvarial defect interfrontal encephalocele causing hypertelorism.

Correspondence
Luis Rafael Moscote-Salazar, RED LATINO
Latin American Trauma & Intensive Neuro-Care Organization, Bogota, Colombia
Email: mineurocirujano@gmail.com

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