

Cloverleaf Skull Syndrome: A Rare Case Report

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Abstract:

Cloverleaf deformity is an extremely rare skull deformity that happens when multiple joints (sutures) between a baby's skull bones begin to fuse too premature. Also known as Kleeblattschädel syndrome, the cloverleaf deformity causes bossing at the front and sides of the skull, resembling a cloverleaf shape. Cloverleaf skull syndrome is an abnormal configuration of the calvaria classified as Craniosynostosis, consisting of premature ossification of cranial bones. It is a deformity characterized by a remarkable enlargement of the head, with a trilobed configuration of the frontal view, resembling a three leaved clover. The first report about such a syndrome in the literature occurred in 1973 and, over the years only some tens of cases have been documented worldwide. The present report describes a case of a severe craniofacial condition that is known as "cloverleaf skull syndrome". On Physical examination multiple malformations were observed, with altered craniofacial configuration, low set ear, hypertelorism, exophthalmos, syndactyly, closed anterior fontanelle, high arched palate, bilateral CTEV. The vast majority of patients with kleeblattschädel have mentalretardation and hydrocephalus. It is usually seen in thanatophoric dysplasia: classically type II only, severe Apert syndrome, severe Crouzon syndrome, Boston-type craniosynostosis & Carpenter syndrome. Craniosynostosis has an overall incidence of 1 in 2100 to 1 in 2500 live births with multi-suture craniosynostosis constituting about 5% of all the cases.

Key Words: Cloverleaf skull, Cloverleaf deformity, Craniosynostosis, Calvaria, Kleeblattschädel syndrome

Introduction:

Cloverleaf deformity is an extremely rare skull deformity that occurs when several joints (sutures) between a baby's skull bones begin to fuse too early. It is also known as Kleeblattschädel syndrome, the cloverleaf deformity causes bulging at the front and sides of the skull, resembling a cloverleaf shape. Babies with cloverleaf deformity often have problems such as: 1) Fluid collecting in the skull 2) Pfeiffer syndrome: Disorder with other conditions including neurological and developmental problems in the fingers and toes 3) Problems breathing because of facial deformities.

Cloverleaf skull syndrome is an abnormal configuration of the calvaria classified as

craniosynostosis, consisting of premature ossification of cranial sutures. It is a deformity characterized by a remarkable enlargement of the head, with a trilobed configuration of the frontal view, resembling a threeleaved clover.[1] This abnormality occurs as a result from a severe alteration in the development of the skull, with premature synostosis of some cranial sutures[1], most commonly the coronal and lambdoid sutures[2], in association with hydrocephalus, leading to a marked bulging of the head in the region of the anterior fontanel and laterally in the temporal regions, with the typical appearance of a "cloverleaf".[1] Syndromic and non-syndromic presentations have been reported.

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ISSN No. : (p) 2348-523X, (o) 2454-1982

DOI: 10.46858/vimshsj.9106

Date of Published : 15th March 2022

Because of the anomalies both in the calvaria and in the skull base and face, this is one of the craniosynostoses currently requiring the most complex multidisciplinary approach.[1] The first report about such a syndrome in the literature occurred in 1973 and, over the years only some tens of cases have been documented worldwide.[3] The present report describes a case of a severe craniofacial condition that is known as "cloverleaf skull syndrome".

Case Report:

A 12 hrs old Full term male child born to primi mother at 38 weeks of gestation via LSCS with birth weight of 3kg referred from civil hospital in view of NICU care. Baby cried immediately after birth. Apgar Score was 7,8,9 at 1,5,10 minutes of life respectively. The baby was stable on room air at delivery but was transferred to neonatal intensive care unit (NICU) for further monitoring. The pregnancy had been uncomplicated, and maternal laboratory test results were negative for HIV, group B streptococci, Syphilis. Fetal ultrasonography not done. There is history of third-degree consanguinity & no history of congenital disorders in her family or in the father's family.

The infant's vital signs had been relatively normal at birth and throughout the NICU stay. Birth weight & length were in normal range for gestational age. At Physical examination multiple malformations were observed, with altered craniofacial configuration, low set ear, hypertelorism, exophthalmos, syndactyly, closed anterior fontanelle, high arched palate, bilateral CTEV. Patient was also having depressed nasal bridge & short neck.(Pic 1) Breath sounds were present; However noisy upper airway sounds were heard. The infant had good range of motion of the Extremities. The rest of the physical examination findings were normal. Heart rate was regular without murmur, bowel sounds were present. The abdomen was soft, with no organomegaly.

On Neurosonogram bilateral dilatation of ventricular system noted. Signs of Closure of saggital, coronal and lamboid sutures were observed. Craniosynostosis was present. The patient was referred to the higher center.



Pic 1: altered craniofacial configuration

Discussion:

It refers to severe type of craniosynostosis which gives skull a cloverleaf shape. It is very rare. It typically results from intrauterine premature closure of the sagittal, coronal and lambdoid sutures, through which the cranial contents bulge, giving rise to the characteristic trilobate shape (hence the name).[4] The vast majority of patients with kleeblattschädel have intellectual disability and hydrocephalus. It is usually seen in the anophthalmic dysplasia: classically type II only, severe Apert syndrome, severe Crouzon syndrome, Boston-type craniosynostosis & Carpenter syndrome.[5] As patient was unaffordable, karyotyping & other tests were not sent.

Fearon et al. conducted a retrospective study of 802 patients treated with craniosynostosis over the past 17 years in their center. Out of the sample they tested, 28 were identified with Pfeiffer's syndrome out of which 17 were classified as type 1 (61%), 7 as type 2 (25%), and 4 as type 3 (16%). The occurrence of hydrocephalus associated with this syndrome was 61%, and they required an intervention to treat the condition.[6] The increased intracranial pressure is manifested as bulging in the areas of the sagittal suture and squamosal sutures with a downward displacement of the ears, severe proptosis or exophthalmos, and facial deformities similar to those of craniofacial dysostosis.[7] The authors identified a 7% mortality rate associated with the syndrome and suggested a reduction in mortality rates with aggressive airway management and more frequent screening for Chiari malformations.[6]

A unique study was done by Witt et al. who screened an 18-gestational-week fetus using standard radiographs and high-resolution CAT scans. The authors reported changes in the calvarium as being the primary focus of abnormal events leading to synostosis. This thought was opposed to the previously established concept that the synostosis was due to cranial base deformities which secondarily distorted the developing brain. This claim was inconsistent with earlier theories regarding the development of craniodyostoses, making it unique.[8]

Craniosynostosis has an overall incidence of 1 in 2100 to 1 in 2500 live births with multi-suture craniosynostosis constituting about 5% of all the cases.[9] This condition is associated with a variety of presentations including optic atrophy, hydrocephalus, respiratory problems, cleft palate, and disorders of hearing and speech.[10] Pfeiffer, Apert, Crouzon, Beare–Stevenson, Jackson–Weiss, and Muenke syndromes are all associated with gain-of-function mutations in the fibroblast growth factor receptor (FGFR) 1, 2, and 3 genes.[9]

According to Machado et al., the diagnosis of cloverleaf skull is based on phenotypical with observation of bitemporal bulging.[11] Cohen's characteristic radiological findings of cloverleaf skull are a tri-lobar skull with distorted calvaria, giving a honeycomb appearance and hypoplasia of orbits along with paranasal sinuses.[2]

The reconstruction of this tri-lobar skull can be done in two in two ways. The first is an early cranial vault remodeling, and the second way is a phased reconstruction, whereby a ventriculoperitoneal shunt is placed in the neonate, fronto-orbital advancement made at 3 to 6 months and posterior vault remodeling done at 1 year of age. The phased reconstruction has shown to have lesser complications in terms of pneumonia, meningitis, excessive bleeding wound infection, seizure, and a shorter intensive care unit and hospital stay as compared to children operated for early cranial vault remodeling.[12] This deformation could lead to hydrocephalus, hindbrain herniation, and venous hypertension. [2]

Conclusion:

Craniosynostosis would lead to a forced growth in perpendicular to the closed sutures. It presents with different syndromes. It leads to gross alteration of skull. It is always with greater risk of morbidity and mortality. These cases are very rare & so shall be reported promptly to deal with management part of this disease. An expert team of maxillofacial, plastic, and neurosurgeons is needed to handle such complex conditions which involve the correction of the bony deformity.

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