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Case Study

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CLOVERLEAF SKULL SYNDROME: A CASE REPORT

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ABSTRACT

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] Until 1981, only 30 cases had been published in the literature, the first of them in 1973, in the ophthalmic literature.[2] The present case report describes severe craniofacial condition that is known as "cloverleaf skull syndrome".

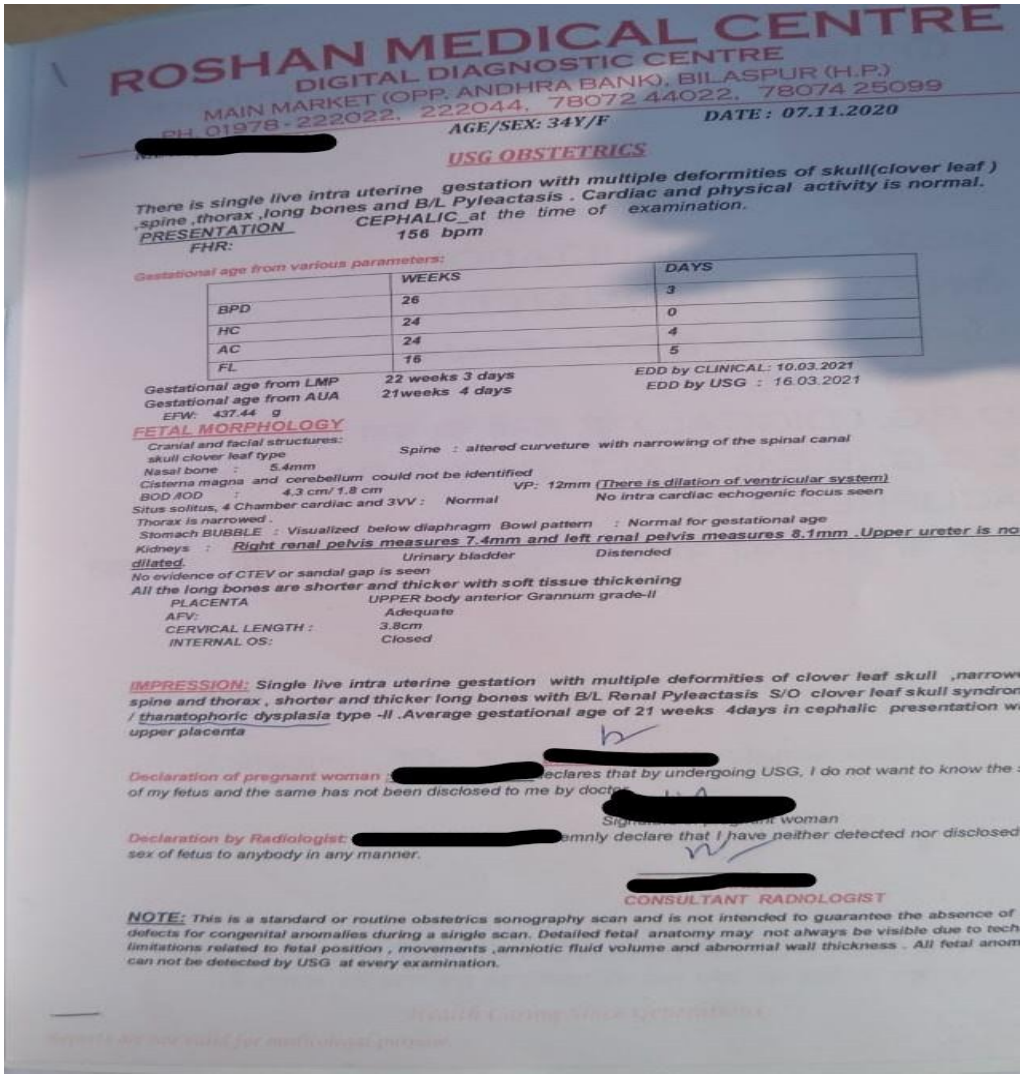
KEYWORDS: Cloverleaf skull syndrome, Craniosynostosis, Thanatophoric dysplasia.

INTRODUCTION

Cloverleaf skull, also known as kleeblattschädel syndrome or deformity, refers to a type of severe craniosynostosis which gives the skull a cloverleaf shape. This abnormality occurs as a result from a severe alteration in the development of the skull, with premature synostosis of some cranial sutures, most commonly the coronal and lambloid sutures, 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] This rare entity is usually diagnosed during the newborn period and is verified by radiological findings. With the advent of ultrasound, and its frequent usage in obtaining bi-parietal diameters, prenatal diagnosis of this skull abnormality is possible.[2]

Case presentation

A 34 year old primigravida who was a booked case at our institution came to open patient door (OPD) at 23 weeks of period of gestation (POG) for routine checkup. The Ultrasound done at 22 weeks of POG shows single live intrauterine gestation with multiple deformities of cloverleaf skull, narrowing of spine and thorax, shorter and thicker long bones with bilateral renal pyleactasis suggestive of cloverleaf skull syndrome/ thanatophoric dysplasis type-II (Figure 1 and 2). The antenatal history was normal except for maternal fever lasting for one months since the third month of gestation, for which the mother took homeopathic treatment. There was no significant family history, and no history of parental consanguinity. Patient was counseled about the outcome of pregnancy and referred to higher institution for further management.



ROSHAN MEDICAL CENTRE
DIGITAL DIAGNOSTIC CENTRE
MAIN MARKET (OPP. ANDHRA BANK), BILASPUR (H.P.)
PH. 01978-222022, 222044, 78072 44022, 78074 25099

AGE/SEX: 34Y/F DATE : 07.11.2020

USG OBSTETRICS

There is single live intra uterine gestation with multiple deformities of skull(clover leaf) ,spine ,thorax ,long bones and B/L Pyleactasis . Cardiac and physical activity is normal.
PRESENTATION CEPHALIC_at the time of examination.
FHR: 156 bpm

Gestational age from various parameters:

	WEEKS	DAYS
BPD	26	3
HC	24	0
AC	24	4
FL	16	5

Gestational age from LMP 22 weeks 3 days EDD by CLINICAL: 10.03.2021
Gestational age from AUA 21weeks 4 days EDD by USG : 16.03.2021
EFW: 437.44 g

FETAL MORPHOLOGY
Cranial and facial structures: skull clover leaf type Spine : altered curveture with narrowing of the spinal canal
Nasal bone : 5.4mm
Cisterna magna and cerebellum could not be identified
BOD AOD : 4.3 cm/ 1.8 cm VP: 12mm (There is dilation of ventricular system)
Situs solitus, 4 Chamber cardiac and 3VV : Normal No intra cardiac echogenic focus seen
Thorax is narrowed .
Stomach BUBBLE : Visualized below diaphragm Bowl pattern : Normal for gestational age
Kidneys : Right renal pelvis measures 7.4mm and left renal pelvis measures 8.1mm .Upper ureter is no dilated. Urinary bladder Distended
No evidence of CTEV or sandal gap is seen
All the long bones are shorter and thicker with soft tissue thickening
PLACENTA UPPER body anterior Grannum grade-II
AFV: Adequate
CERVICAL LENGTH : 3.8cm
INTERNAL OS: Closed

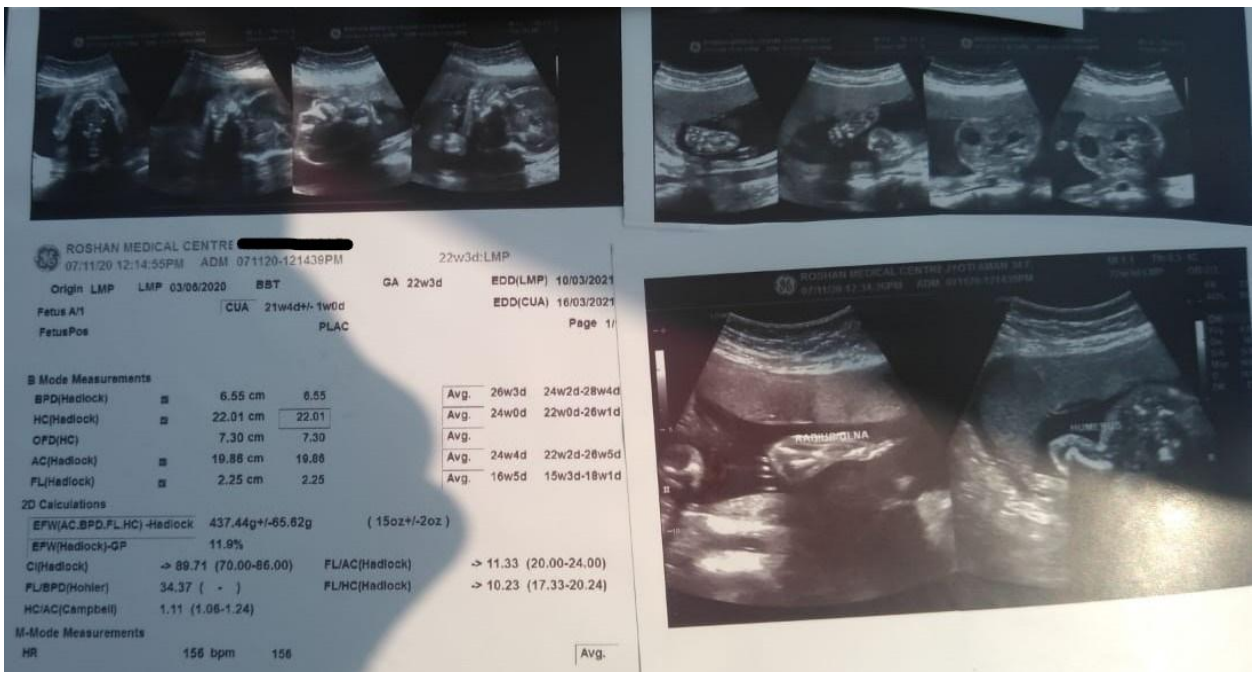
IMPRESSION: Single live intra uterine gestation with multiple deformities of clover leaf skull ,narrow spine and thorax , shorter and thicker long bones with B/L Renal Pyleactasis S/O clover leaf skull syndrom / thanatophoric dysplasia type -II .Average gestational age of 21 weeks 4days in cephalic presentation wi upper placenta

Declaration of pregnant woman : [redacted] declares that by undergoing USG, I do not want to know the s of my fetus and the same has not been disclosed to me by doctor. [redacted]
Signature of pregnant woman
Declaration by Radiologist: [redacted] solemnly declare that I have neither detected nor disclosed sex of fetus to anybody in any manner.

[redacted]
CONSULTANT RADIOLOGIST

NOTE: This is a standard or routine obstetrics sonography scan and is not intended to guarantee the absence of defects for congenital anomalies during a single scan. Detailed fetal anatomy may not always be visible due to tech limitations related to fetal position , movements ,amniotic fluid volume and abnormal wall thickness . All fetal anom can not be detected by USG at every examination.

Figure 1: Prenatal ultrasonography of cloverleaf skull syndrome.



ROSHAN MEDICAL CENTRE [redacted]
07/11/20 12:14:55PM ADM 071120-121439PM
Origin LMP LMP 03/06/2020 BBT
Fetus A/1 CUA 21w4d+/- 1w0d
FetusPos PLAC
22w3d:LMP
GA 22w3d EDD(LMP) 10/03/2021
EDD(CUA) 16/03/2021
Page 1/

B Mode Measurements

BPD(Hadlock)	6.55 cm	6.55	Avg.	26w3d	24w2d-28w4d
HC(Hadlock)	22.01 cm	22.01	Avg.	24w0d	22w0d-26w1d
OFD(HC)	7.30 cm	7.30	Avg.		
AC(Hadlock)	19.86 cm	19.86	Avg.	24w4d	22w2d-26w5d
FL(Hadlock)	2.25 cm	2.25	Avg.	16w5d	15w3d-18w1d

2D Calculations

EFW(AC,BPD,FL,HC)-Hadlock	437.44g+/-65.62g	(15oz+/-2oz)	
EFW(Hadlock)-GP	11.9%		
CI(Hadlock)	-> 89.71 (70.00-86.00)	FL/AC(Hadlock)	-> 11.33 (20.00-24.00)
FL/BPD(Hohler)	34.37 (-)	FL/HC(Hadlock)	-> 10.23 (17.33-20.24)
HC/AC(Campbell)	1.11 (1.06-1.24)		

M-Mode Measurements

HR	156 bpm	156	Avg.

Figure 2: Prenatal diagnosis of cloverleaf skull.

DISCUSSION

Cloverleaf skull syndrome is a rare presentation of craniosynostosis with clinical features consisting in trilobed skull, exophthalmos, low ear implantation and upper airway. Hydrocephalus is also a common finding, despite the multifactorial nature of such an abnormality.[3] It occurs as a result from a severe alteration in the development of the skull, with premature synostosis of some cranial sutures, most commonly the coronal and lambloid sutures, 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.[1-2] Hypoplasia of maxilla, nasal, zygomatic bones and orbits, high-arched palate, and micromelia with chondrodystrophic changes may also occur. Most children are still-born or die soon after birth without mental development.[2]

The precise etiopathogenesis of this syndrome is still to be completely known, with theories involving altered membranous-osseous and/or endochondral ossification, generalized chondrodysplastic process, and a possible vascular origin associated with the abnormal osteoclastic resorption.[4] The diagnosis of such a syndrome can be made in the prenatal period by means of ultrasonography, which detects the altered cranial morphology and hydrocephalus. Traditionally, the diagnosis occurs during routine prenatal follow up at the second gestational trimester. However, with the increasing use of obstetric ultrasonography at the first gestational trimester, such alterations may be detected increasingly earlier over the gestation.[5] The majority of sonographic case studies involving cloverleaf skull associated with thanatophoric dysplasia and hydrocephaly[5] as seen in present case study. Prenatal diagnosis allows the parents to make an informed choice about continuation of pregnancy. It is noted that reported cases of surgical management of this entity are very rare. Total craniectomy proved to be a satisfactory treatment, one which was responsible for the reversal of hydrocephalus.[6] Early detection and treatment of this rare malformation results in acceptable cosmetic and neurologic improvement in some patients, yet in developing countries like ours parents should be counseled about outcome of pregnancy and their decision for continuation or termination of pregnancy should be respected.

CONCLUSION

In cloverleaf skull syndrome most children are still-born or die soon after birth without mental development. Early diagnosis of this rare disorder allows parents to make an informed choice for continuation of pregnancy and its outcome.

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