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A Rare Case of Arnold Chiari – 2 Anomaly

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Abstract:

➢ ***Introduction:***

Arnold–Chiari malformations represent structural defects of the hindbrain, first characterized by Austrian pathologist Hans Chiari in the late 19th century. These anomalies involve the posterior cranial fossa and its neural structures, including the cerebellum, pons, and medulla oblongata. Type 2 Chiari malformation is a relatively frequent congenital defect characterized by a beaked midbrain, caudal displacement of the cerebellar tonsils and vermis, and an associated spinal myelomeningocele.

➢ ***Case Report:***

We describe an unbooked 20-year-old primigravida presenting at approximately 6 months of amenorrhea with decreased fetal movements and abdominal pain. Antenatal ultrasound revealed lumbosacral spina bifida with features consistent with Arnold–Chiari type 2 malformation. Following counseling, pregnancy termination was performed, and the fetus demonstrated myelomeningocele with hydrocephalus on gross examination.

Keywords: *Arnold–Chiari Malformation; Spina Bifida; Primigravida; Congenital Anomaly; Prenatal Diagnosis.*

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I. INTRODUCTION

Arnold–Chiari malformations are a spectrum of congenital hindbrain anomalies resulting from defective embryologic development of the posterior fossa. First described by Hans Chiari in the 1890s, these malformations produce a range of neurological and structural manifestations, from herniation of the cerebellar tonsils to complete cerebellar absence. They may occur alone or with other intracranial or spinal abnormalities such as hydrocephalus, encephalocele, syringomyelia, and spinal dysraphism.

➢ *Classification:*

- *Type I:*

Elongated, pointed cerebellar tonsils extending >5 mm below the foramen magnum.

- *Type II:*

Caudal displacement of the cerebellar vermis, medulla oblongata, and fourth ventricle, commonly with myelomeningocele.

- *Type III*

Downward migration of all posterior fossa contents into a high cervical or suboccipital encephalomeningocele.

- *Type IV:*

Severe cerebellar hypoplasia with medullary ectopia.

➢ *Epidemiology:*

Type I is most common (0.5–3.5% prevalence), with a slight female predominance. Type II occurs in approximately 0.44 per 1,000 live births, with no gender bias, while Types III and IV are rare.

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➢ *Prognosis:*

Type I generally has a favorable outcome. In contrast, Type II is associated with significant morbidity and mortality, with neonatal in-hospital mortality of ~3% and a 3-year mortality rate of ~15%.

II. CASE REPORT:

A unbooked case of 20yrs old patient primi with 6 months of ammenorhea came with complaints of decreased perception of fetal moments along with pain abdomen which is insidious in onset , non progressive, spasmodic type & non radiating to back since 15 days was admitted on 22/10/23.

General physical examination and systemic examination was uneventful.

Per abdomen examination- uterus corresponding to 24-26 weeks period of gestation, relaxed ,FHS heard on auscultation,external ballotment present.

ANC profile and USG was done

➢ *Investigations*

CBC: Hb-8.7gm%, TC- 12000cells, Blood grouping and typing- AB positive, urine routine – Normal, Coagulation profile- Normal, Serology- Non reactive, RBS- 84mg/dl, TSH-4.8microgram/dl, Peripheral smear – Microcytic hypochromic anemia.

➢ *Usg Anomaly Scan –*

Single live intrauterine fetus of 27 weeks + 2d period of gestation with placenta anterior grade 1 , FHR- 155bpm, EFW-700gms , AFI- Adequate, variable presentation

Fetal brain & face – skull appears lemon shaped with bilateral lateral ventricles are mildly dilated measuring 9mm at maximum width suggestive of ventriculomegaly with absent cerebellum & corpus callosum noted .Fetal spine – there is significant splaying of posterior elements at lumbosacral region noted suggestive of spina bifida.

Findings likely represent Type- 2 Arnold Chiari malformation

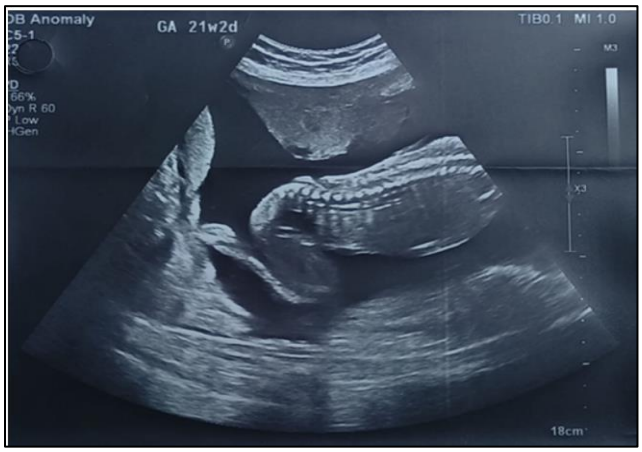

Fig 1 Absence of Spinous Process & Posterior Vertebral Body. Significant Widening of Lumbo Sacral Region

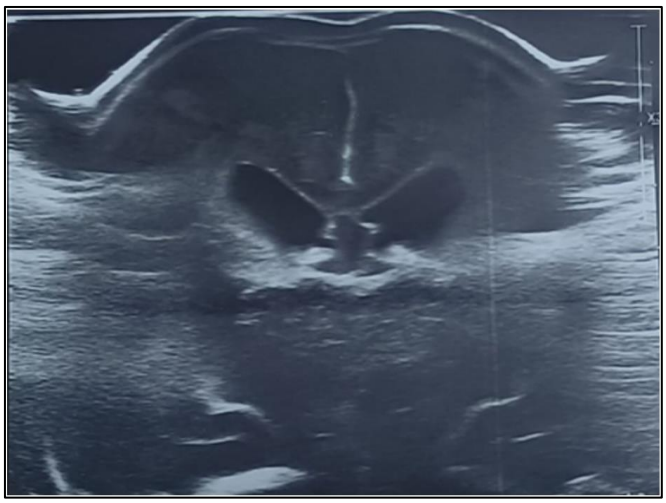

Fig 2 Absence of Cerebellum & Corpus Callosum (Banana Sign)

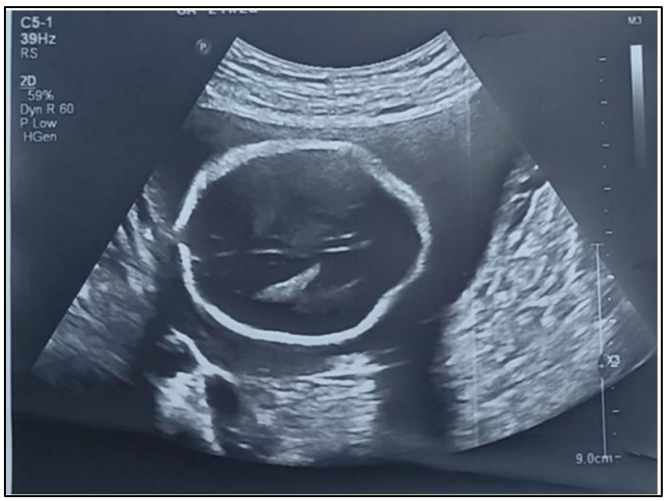

Fig 3 Lemon Shaped Skull

➢ *Management*

- Patient was explained about the congenital anomaly & the need for termination of pregnancy & consent was taken.
- One pint PRBC was transfused before the procedure.

Tab. Mifepristone 200mg P/O given and labour is induced with foleys induction and Injection oxytocin 10 IU in ringer lactate fluid IV was started. Delivered a single live female fetus with placenta intoto, of birth weight 700mg English units as identifiers in trade, such as “3.5-inch disk drive.”

➢ *On Gross Examination*

The baby had hydrocephalus with Open spinal cord with meningomyelocele is present in the lumbosacral region suggestive of spina bifida.

Infantogram was done ,similar findings were noted.

Karyotyping or fetal autopsy could not be conducted due to patient refusal

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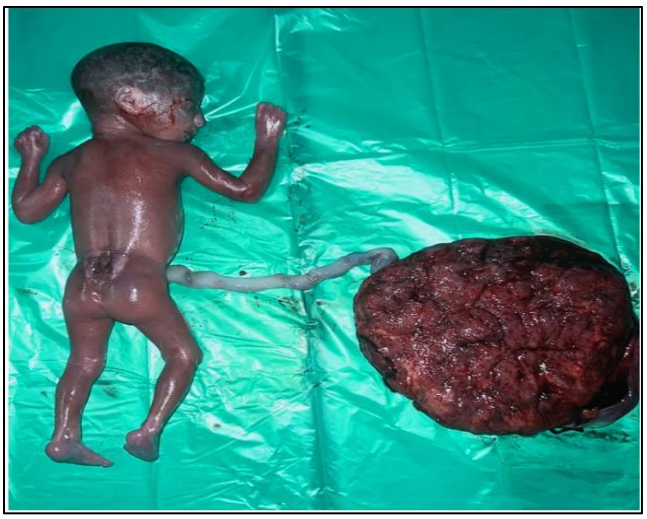

Fig 4 Images Showing Spina Bifida

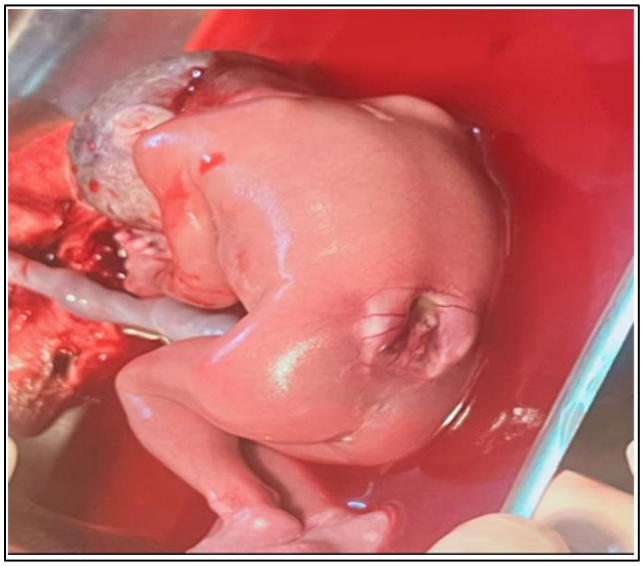

Fig 5 Showing Spina Bifida

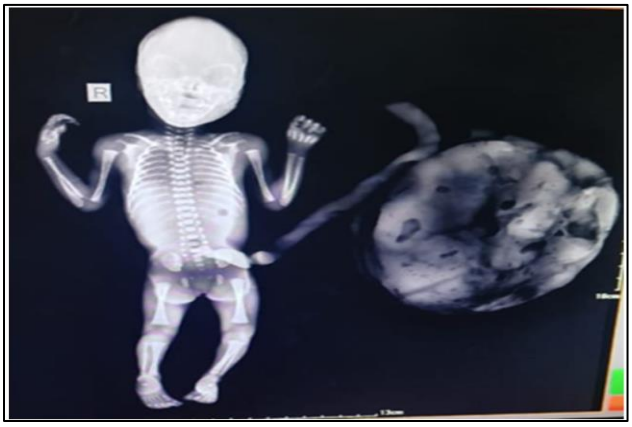

Fig 6 Showing Infantogram of the Fetus

III. DISCUSSION

Chiari Type 2 malformation is almost universally associated with thoracolumbar or lumbosacral myelomeningocele. The hallmark features include downward herniation of the cerebellar vermis, medulla, and fourth ventricle into the spinal canal. Hydrocephalus develops in approximately one-quarter of cases, often due to aqueductal stenosis or obstruction of the fourth ventricular outflow.

Radiological indicators such as the lemon sign (scalloping of the frontal bones) and banana sign (curved cerebellum with effacement of the cisterna magna) are highly suggestive of this malformation and aid in prenatal detection. In this case, both signs were present, along with ventriculomegaly and spinal dysraphism.

Prenatal detection facilitates early counseling, consideration of pregnancy termination, and prevention of maternal and neonatal complications. While surgical repair of myelomeningocele is possible postnatally, outcomes are variable, and neurological deficits often persist.

IV. CONCLUSION

All women planning to conceive should receive daily intake of 400 micrograms of folic acid in the pre conception period & to be continued for 3 months in pregnancy.

All pregnant women are advised for regular antenatal check up with NT scan between 11-13+ 6 days weeks & anomaly scan between 18-24 weeks is advised.

Early diagnosis of neural tube defects helps to make decision about termination of pregnancy to prevent maternal complications and neonatal morbidity.

Genetic counselling also to be done.

Congenital anomalies can be prevented by folic acid supplementation.

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