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Arnold Chiari Malformation With Meningo-Myelocele And Open Spina Bifida For MTP: A Case Report

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ABSTRACT

In this case report we report a 39-year-old primigravida with 20 weeks+3 days presenting with abortion. She presented to the antenatal clinic with Amenorrhea since 5 months and bleeding per vagina since 1 day. Her ultrasonography report showed that single live intrauterine gestation of 20 weeks 3 days, fetal cardiac activity presented, liquor was adequate, placenta anteriorly implanted, fetal cranium appeared abnormal with frontal bone flattening with lemon shaped appearance and small posterior cranial fossa. Cerebellum appears banana shaped. She was diagnosed to have Arnold chiari malformation and meningomyelocele with open spina bifida in second trimester and medical termination of pregnancy was carried out.

Keywords: Arnold Chiari Malformation, Spina Bifida, Classication, Diagnosis And Management

I. Introduction

A complex clinical and pathological disorder including malformation of the cerebellum and brainstem in children was documented by pathologists Julius Arnold (1835-1915) and Hans Chiari (1851-1916) near the end of the nineteenth century. According to current definitions, Arnold Chiari malformations are a range of anomalies in the hindbrain that affect the brainstem, cerebellum, cervical cord, and base of the skull. Four categories of Chiari malformations have been established based on the brain or spine development abnormalities and the type of brain tissue herniation that has been displaced in the spinal canal[1]

In Chiari Malformation Type I (CM-I), the cerebellar tonsils herniate beneath the foramen magnum. The more complicated Chiari Malformation Type II (CM-II), which is the subject of this report, involves the descent of the brainstem, vermis, fourth ventricle, and cerebellar tonsils. Chiari Malformation Type IV (CM-IV) is an uncommon disorder linked to an undeveloped cerebellum, while Chiari Malformation Type III (CM-III) is defined by a high cervical or low occipital encephalocele [3]. There is a 0.44 out of 1,000 infants with a Chiari II malformation, with no gender preponderance. Supplementing with folate during pregnancy can reduce the occurrence of myelomeningocele because of its correlation with it.

The "unified theory," which maintains that the cranial traits are a result of the neural tube abnormality, is the most modern and widely recognized theory. Previous theories based on the myelomeningocele tying the spinal cord and causing caudal tension on the brainstem and hindbrain regions have lost support.Myelomeningocele is almost usually linked to the complicated congenital brain malformation known as Chiari type II malformation (Arnold-Chiari malformation), which is the most prevalent severe deformity of the posterior fossa [2].

Chiari II malformation can cause a number of clinical symptoms, such as brainstem symptoms, tethered cord syndrome or syringomyelia, spinal symptoms from myelomeningocele, subsequent hydrocephalus, or lower cranial nerve dysfunction. Fetal ultrasonography can sometimes identify Chiari II malformation. The cerebellar vermis, brainstem, and fourth ventricle caudal displacement are the hallmarks, although there may also be the following findings: The frontal bone indentation that resembles a lemon is called a "lemon sign." The sign for the banana cerebellum indicates how the cerebellum encircles the brain stem. Beaking of the tectum a myelomeningocele.

The degree of the craniospinal deformity and related neurological deficits determine how to treat Chiari II malformation. Patients with Chiari 2 malformation have a very serious natural history, and medullary symptoms continue to be the primary cause of death. Repairing related open neural tube problems frequently involves surgery. In order to repair myelomeningocele, surgery is frequently necessary. The advantages of undergoing myelomeningocele repair on the fetus while it is still in the womb have been shown by some evidence. Delivery should preferably take place in a facility with a level III neonatal intensive care unit for infants diagnosed with myelomeningocele who did not have a fetal intervention performed. After birth, the first course of treatment

consists of covering the lesion and giving preventative antibiotics. It is recommended that the open neural tube defect be surgically corrected during the first 72 hours[1].

Case Report

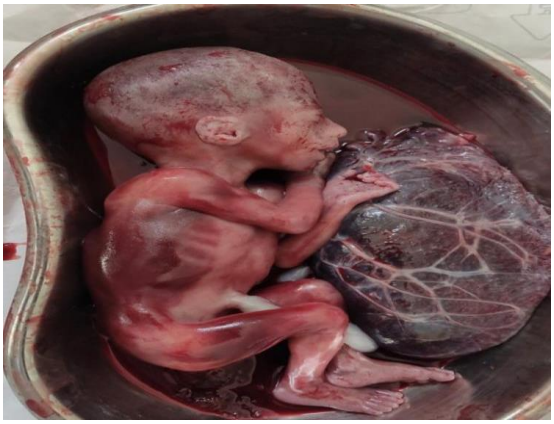
Mrs X, 39 years old primigravida presented to the antenatal clinic with chief complaints of 5 months Amenorrhea and bleeding per vagina since one day. Her menstrual cycles were regular. She gave no history of diarrhea, constipation, fever, urinary complaints and recent illness. She conceived spontaneously. She detected her pregnancy after 14 days of missed period. She had regular antenatal visit. She took two doses of tetanus. No past history of any kind of disease like Diabetes Mellitus, Hypertention, Tuberculosis, Asthma, Epilepsy, any Congenital malformation and blood transfusion. She had taken vaccination of COVID-19(covisheild) in October, 2021.

On examination patient was calm, conscious, oriented and coherent with pulse 84 bpm, blood pressure 112/70mmHg, temperature afebrile, cardiovascular, central nervous system and respiratory systems normal. Abdominal examination revealed fundal height corresponding to 20 weeks gestation. Uterus was relaxed, non tender and external ballottment presented. There was a single fetus in longitudinal lie. Fetal heart rate was good and regular. On vaginal examination Os parous and uneffaced.

All her blood components which included Prothrombin time, hemoglobin, Eosinophils, MCHC were decreased, polymorphs increased and urine investigations was within normal limits. Her ultrasonography reported that single live intrauterine gestation of 20 weeks 3 days, fetal cardiac activity presented, liquor was adequate, placenta anteriorly implanted, fetal cranium appeared abnormal with frontal bone flattening with lemon shaped appearance and small posterior cranial fossa. Cerebellum appears banana shaped. There was dilatation of lateral ventricles measuring ~12mm. In fetal spine, there was splaying of posterior elements in the lumbo-sacral region with a small defect, through which hypo to anechoic content were seen herniating, measuring ~19X18mm. The findings of biometry- Biparietal Diameter 19 weeks+5 days, Head Circumference- 21 weeks, Abdominal Circumference- 20 weeks+ 2 days, Femur length- 20 weeks + 3 days, Expected fetal weight- 347 gm. The findings were suggestive of Arnold chiari II malformation with meningo-myelocele and open spina bifida in sacral region. The plan of treatment was medical termination of pregnancy in second trimester.

In medical intervention, the labor is induced by giving the following drugs for the expulsion of the conception through the vagina. Tab. Misoprostol 400mg P\V STAT, RL fluid with Inj. Oxytocin 10 IU in 500ml I\V STAT, Inj. oxytocin 10 IU I\M STAT, Inj. Voveran 75mg I\M STAT, Inj. Ceftriaxone 1gm I\V STAT, Tab. Cabergoline 0.25mg P\O STAT, Tab. Meftalspas BD P\O STAT.

Under all aseptic precautions, patient cleaned and draped. Patient in lithotomy position, delivered a live female fetus, weigh 330gm, length 22cm, head circumference 18cm, abdominal circumference 14cm, bilateral lower limbs 11cm with 5 finger in both feet and hands. Fluid filled with defect of 222cm on sacral region. Placenta delivered spontaneously, 5 min. following delivery of fetus, complete with membrane, weigh 148 gm, discoidal with eccentric, cord length 35cm, no gross, CNF seen.

According to the physician instructions, the patient should drink a lot of water, eat a diet heavy in iron and protein, and practice good personal hygiene. psychological and emotional assistance given to the customer. In addition to counseling on the use of family planning techniques to prevent an unintended pregnancy and condoms to prevent infection with STIs, HIV, and AIDS, it is advised that a minimum of six months should pass between a spontaneous abortion and the next pregnancy for the sake of the mother's and the unborn child's health, as well as reproductive and other health services, such as nutritional screening and treatment of nutritional deficiencies, hygiene education, prevention of further danger signs, and follow-up.

Discussion

ACM II, or Arnold Chiari Malformation Type II, is a complicated and uncommon neurological condition that is commonly referred to as Chiari II malformation. Structural abnormalities in the brainstem and cerebellum, where portions of both protrude into the spinal canal, are the hallmark of this disorder, which is frequently identified in infancy or early childhood. Although the precise prevalence of ACM II is difficult to determine, myelomeningocele, a kind of spina bifida, is intimately linked to it. This correlation is found in roughly 90% to 95% of myelomeningocele patients. Worldwide, there are variations in the prevalence of spina bifida and, consequently, ACM II, but it is thought to impact roughly 1 in 1000 live births[3].

There are currently the following primary causes behind the Arnold-Chiari anomaly's development: violation of the central nervous system's (CNS) embryonic development combined with a skull and spine abnormality. The development of types II and III Chiari malformations is mostly influenced by this component. Narrowing may result from craniosynostosis, or inadequate growth of the skull, particularly in the posterior fossa. The cerebellum, which is forced downward through the foramen magnum, is thought to be compressed as a result. One of the causes of Chiari malformation type II is spinal cord damage. It is brought on by a congenital spinal hernia, typically in the lumbosacral area. Neural processes in the posterior fossa are displaced southward through the foramen magnum as a result of the accompanying spinal cord stretching during the spine's longitudinal expansion. CSF dynamics are broken.
The following neurological syndromes, including hypertensive-hydrocephalic syndrome, cerebellar syndrome, bulbar pyramidal syndrome, radicular syndrome, and vertebrobasilar insufficiency syndrome, can be associated with the Arnold-Chiari anomaly. The brain's penetration into the spinal cord's subarachnoid space causes hypertensive-hydrocephalic syndrome. It has nowhere to drain (except from the normal mechanisms of absorption, which in this instance are insufficient) and the brain's vascular plexuses continue. As CSF builds up in the brain, it can lead to hydrocephalus, which is an expansion of the compartments that store liquid, and intracranial hypertension, which is an increase in intracranial pressure.

Movement coordination problems, a "drunken" walk, and a brief fall when making intentional motions are all signs of cerebellar syndrome. Patients are concerned about lightheadedness. The limbs may tremble. Speech impairment might include chanting or splitting speech into distinct syllables. "Downward nystagmus" is a rather distinctive symptom. These are involuntary eye movements, specifically downward ones. Patients who have nystagmus may complain of double vision.
The compressed structures are what give rise to the label "bulbar pyramidal syndrome." Since the medulla oblongata is known by the name Bulbus, bulbar syndrome denotes symptoms of medulla oblongata injury. The medulla oblongata, which are bundles of nerve fibers that transmit impulses from the cerebral cortex to the anterior horns of the spinal cord, is anatomically represented by the pyramids. The trunk and limbs' voluntary movements are controlled by the pyramids. As stated above, bulbar pyramidal syndrome is clinically characterized by limb muscular weakness, numbness, lack of pain, and sensitivity to cold. When the brain stem's cranial nerve nuclei are compressed, it results in visual and auditory impairments, speaking problems (from a lack of tongue movement), nasal voice, choking during meals, and breathing difficulties. Loss of muscular tone with retained consciousness or a brief loss of consciousness are both possible. In cases of Arnold-Chiari abnormality, radicular syndrome manifests as symptoms of cranial nerve dysfunction. These include decreased tongue mobility, a hoarse or nasal voice, difficulty swallowing food, hearing impairments (including tinnitus), and decreased facial sensibility.

Impaired blood flow in the relevant blood pool is linked to vertebrobasilar insufficiency syndrome. As a result, dizzy attacks, loss of awareness or muscular tone, and vision issues occur. As you can see, the majority of Arnold-Chiari malformation symptoms are not caused by a single direct cause, but rather by a combination of factors, including damaged blood flow in the vertebrobasilar basin and compression of particular medulla oblangata centers, which can result in attacks of unconsciousness. Similar circumstances arise with hearing loss, vision impairment, vertigo, and other conditions. Syring myelitis syndrome only happens when there is a confluence of spinal cord cystic abnormalities and Arnold-Chiari malformation. These conditions include numbness and muscle weakness in certain limbs, pelvic organ dysfunction (urinary and fecal incontinence), and dissociated sensory disturbances (when temperature, pain, and tactile sensitivity are isolated in isolation and deep (limb position in space) remains intact)[4].

The gold standard for diagnosing congenital abnormalities in utero is ultrasound examination. According to reports, combining ultrasound and biochemical screening can increase the detection rate of open spina bifida in the first trimester by as much as 80–90%. Before the 12th postmenstrual week, myelomeningocele can be identified on fetal ultrasonography by abnormalities of the spine or a bulging of the posterior contour of the fetal back. Various authors claim that there is a soft marker for NTD in the first trimester. The fourth ventricle, which is regarded as Intracranial Translucency (IT) between weeks 11 and 13 + 6 of pregnancy, is this sign. It has been

determined that a lack of IT is a sign of spina bifida.To differentiate between open and closed spina bifida, the amount of alpha-feto protein (AFP) in amniotic fluid can be useful. The maternal serum alpha fetoprotein test, which has a sensitivity of 85% for identifying neural tube abnormalities, is commonly conducted between weeks 15 and 20–22 of pregnancy. It has a threshold of 2.5 multiples of the median value. While AFP concentrations in closed defects are always within normal levels, they are nearly always elevated in open defects. It has been discovered that the amount of acetylcholinesterase in amniotic fluid is more specific than AFP and could be helpful in differentiating between open and closed abnormalities.

Despite the fact that the spinal defect can be identified early in pregnancy, open spina bifida is usually diagnosed in the middle of the second trimester. Since the prenatal detection rate of meningomyelocele is between 86 and 96 percent, the mid-1980s saw a considerable improvement in the prenatal diagnosis of this condition by reporting a number of distinctive intracranial abnormalities that were simpler to establish than direct imaging of the spinal defect. Both longitudinally and transversely, the neural tube from the cervical to the sacral area must be thoroughly examined. The typical spine appears as a complete circle with intact skin when viewed in cross-section. The injured vertebral arch in spina bifida aperta cases gives it a U shape, which permits the spinal cord to protrude (meningocele or myelomeningocele). The cerebellar vermis, fourth ventricle, and medulla oblongata are displaced through the foramen magnum due to a leak of cerebrospinal fluid into the amniotic cavity (Arnold-Chiari or Chiari II malformation), resulting in pathognomonic cerebral abnormalities that are detectable by ultrasound. Changes in the posterior cerebral fossa are the most sensitive and specific finding for the diagnosis of spina bifida. The "banana sign" on an axial section sonogram is a representation of the ruptured cerebellum in the foramen magnum. A smaller cisterna magna and a smaller cerebellum—defined as a transcerebellar diameter below the 10th percentile for the gestational age—are the causes of the abnormality. The "lemon sign," which is typically seen, is a description of the skull's shape in the transverse plane of scanning that is defined by the concavity of the frontal bones close to the coronary sutures. According to the findings, the afflicted fetuses were identified as early as 13 weeks and in 50–90% of cases before to 24 weeks of gestation, but only 13% after this time. It should be noted that up to 1% of normal fetuses exhibit a small concavity of the frontal bones, making the lemon sign non-pathognomonic for meningomyelocele. It is also feasible to "make" a lemon sign if the skull is not shown in the proper axial plane.

Prenatal open/fetoscopic surgery, pregnancy termination, or continuing the pregnancy with postnatal surgical repair are the three primary options available to the practitioner and the family upon diagnosis of spina bifida aperta. In the United States, myelomeningocele has been surgically corrected during the first 24 weeks of pregnancy since 1994. It is thought that prenatal correction will preserve motor activity more than postnatal surgery, even if brain capabilities cannot be restored with this therapy. Despite these positive outcomes, a 30% chance of complications was noted, including early birth, placental abruption, early fetal mortality, uterine rupture with intra-abdominal hemorrhage, and the ensuing threat to the mother's life.

Although the long-term results are yet unknown and the rate of dehiscence and leaking from the myelomeningocele repair site can be higher, percutaneous fetoscopic repair is probably associated with fewer serious maternal problems. Many would-be moms in the United States of America today decide to end their pregnancies rather than face the possible risks; this is also the case in Bulgaria, where fetal surgical repair is still a possibility for the future[5].

Surgery to restore cerebrospinal fluid flow via the craniovertebral junction and decompress the posterior fossa to reduce pressure on the cerebellum and hindbrain is the primary treatment for Chiari malformation. Patients with persistent hernia symptoms are advised to have surgery. If symptoms do not appear for asymptomatic tonsil hernias, follow-up is advised. Surgery that is done within two years after the onset of symptoms yields the greatest surgical outcomes.

The myelomeningocele is corrected surgically as the first step in treating Chiari II, typically within the first 48 hours. Additionally, a hysterotomy can be performed in utero. There are several methods for closing spinal dysraphism, including musculocutaneous flaps and primary skin closure, depending on the severity, layers involved, and available nearby tissue. To drain the cerebrospinal fluid in the event of hydrocephalus, the great majority will eventually require a ventricular shunt. In order to permit suboccipital extension, posterior decompression is carried out later if required [4].

If a mother decides to carry the pregnancy to term, she will have to prepare for a child that will require a lot of care and have expensive medical bills. Nearly 14% of all spina bifida newborns die before turning five years old, even with intensive intervention; the death rate increases to 35% in those who have signs of brainstem dysfunction brought on by the Arnold-Chiari malformation. Even with modified accommodations, only half of patients can live freely as adults, despite the fact that 70% of them have an I.Q. above 80.

In our instance, the pregnancy was ended for health reasons. After genetic analysis of the abortive material, no genetic abnormalities were discovered. No evidence of teratogenic drug use, diabetes, or obesity was found. Defects in the neural tube appear by day 35 of embryonic development. This is why, in certain situations, folic acid consumption alone is insufficient to identify pregnancy. About 75% of the risk of spina bifida is decreased when 400 mcg of folic acid per day is added to the diet for three months prior to conception and during the first trimester.

As a final consideration, in many regions of the world, mothers of fetuses with myelomeningocele still have the legitimate choice of terminating their pregnancy. There are ethical issues with this matter by nature, and religious considerations in the area are crucial. For instance, for the past 20 years, women in Western Europe who have a fetus with the illness detected antenatally have the option to terminate their pregnancy at any gestational age. As long as the procedure is completed prior to the third trimester, the option is currently present in the United States of America. In South Africa, regardless of the gestational age, it is legally allowed to end a pregnancy if the fetus has been diagnosed with myelomeningocele[5].

Conclusion

Arnold Chiari Malformation Type II is a complicated disorder that primarily affects newborns, particularly those who also have myelomeningocele. For those who are impacted, treating the illness and enhancing their quality of life depend heavily on early diagnosis and management. The use of CT and MRI imaging is essential for a precise diagnosis. Research must continue in order to better understand the cause, enhance diagnostic techniques, and create more potent treatment plans.

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