Case Report

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Meningomyelocele with Hydrocephalus: A Case Report

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Abstract

Neural tube defect (NTD) is a complex congenital anomaly affecting meninges, vertebral arches, muscle and skin. Spina bifida denotes non-fusion of two vertebral arches causing herniation of meninges and spinal cord through it which is termed as meningomyelocoele (MMC). Meningomeylocoele develops around 4th week of gestation and accounts for an incidence of 1 per 1000 live births. Nearly every case of MMC is associated with hydrocephalus. These neural tube defects form a greater part of fetal morbidity and mortality. Early surgery should be performed because of risk of infection and to prevent further damage to nervous tissue. Proper medical and nutritional advice can reduce the incidence of neural tube defects.

Keywords: Neural Tube Defect; Meningomyelocoele; Hydrocephalus.

Introduction

Spina bifida is a developmental anomaly that occurs when vertebral arches fail to fuse and thus creates a defect. Herniation of spinal cord and meninges occurs through this defect known as meningomyelocoele. Spina bifida develops in 4th week of gestation [1].

Meningomyelocoele is the most common form of neural tube defects [2] with incidence of approximately 1 in 1000 live births[3]. Other congenital conditions associated with spina bifida with meningomyelocoele are hydrocephalus, club foot, dislocation of hip, exstrophy of bladder and rarely cardiac defects. Amongst them hydrocephalus is most common presentation [4]. We present a case of a day old neonate who presented with spina bifida with meningomyelo-coele with hydrocephalus because of its rarity.

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Case Report

A one day old full term female baby was born to a 20 year old gravida-1 para-1 mother by caesarean section. The mother had no H/O drug intake, radiation exposure, or any illness during pregnancy. Mother never attended ANC visits and never received folic acid tablets or supplements.

Birth wt of baby was 3.04 kg with APGAR score 7. On physical examination the baby had enlarged head, having circumference of 41.5cm (Figure 1) and a cystic swelling at lumbosacral region of size 6x4cm with ruptured membranes over it (Figure 2). There was no pallor, icterus, cyanosis, clubbing, oedema or lymphadenopathy. Ophthalmic examination showed positive sunset sign (Figure 3).

On central nervous system (CNS) examination patient was conscious and active but both lower limbs showed hypotonia and absence of reflexes. Other examinations like cardiovascular system, respiratory system and per abdominal examination were normal.

Ultrasonography (USG) skull showed enlargement of both lateral ventricles and dilatation of 3rd& 4th ventricles. USG local part showed evidence of 1.2 cm defect in lumbosacral region with herniation of contents of spinal cord suggestive of meningomyelocoele. Considering this clinical profile

ventriculoperitoneal shunt was done for hydrocephalus & surgical closure was planned for meningomyelocoele afterwards.

Fig. 1: Hydrocephalus-enlarged head, having circumference of 41.5cm

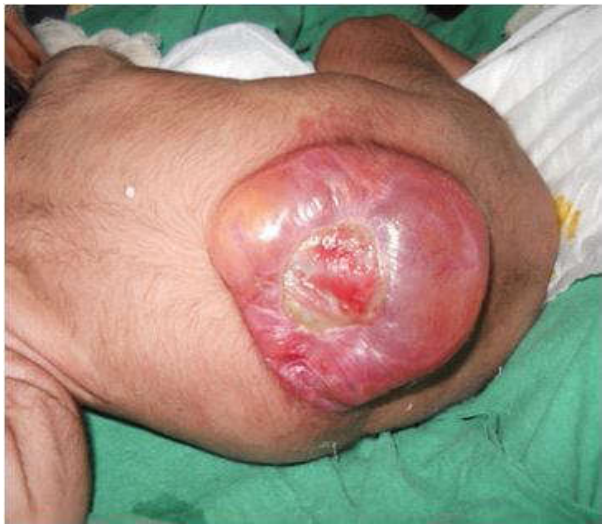

Fig. 2: A cystic swelling at lumbosacral region of size 6x4cm with ruptured membranes over it

Fig. 3: Positive sunset sign

Discussion

Development of CNS involves series of events. By 20th day of gestation neurulation i.e. formation of neural plate, neural fold and its closure to form neural tube starts. First the surface ectoderm of trilaminar embryonic disc gets thickened to form the neural plate. This neural plate invaginates on its central axis to form median neural groove having neural folds on either side. These neural folds fuse to form neural tube and it gets detached from surface ectoderm. Notochordal process is the precursor of spine. Mesoderm gathers around notochord to form the primitive spinal column. Further it gets segmented and form vertebral bodies. Mesoderm condenses over dorsum of spinal cord to form neural arch [5].

Severe neural tube defects involving neural and non-neural structures can occur at any developmental stage. The incidence varies approximately 1/1000 births to 1/100 births depending on the population [3]. Meningomyelocoele is hernial protrusion of meninges plus neural tissue resulting from congenital failure of neural tube to close [6]. Spina bifida is a general term for NTDs with multifactorial etiology, affecting the spinal region consists of a splitting of the vertebral arches & may or may not involve underlying neural tissue. There are two different types of spina bifida [7] .

A. *Spina Bifida Occulta*: It accounts for 10% of cases.

- This defect is due to a lack of fusion of the vertebral arches.
- It occurs in the lumbosacral region.
- It is covered by skin, does not involve underlying neural tissue & marked by a patch of hair overlying the affected region.

B. *Spina bifida cystica*: It is a severe type NTD.

- Neural tissue and/or meninges along with skin protrude through a defect in the vertebral arches to form a cyst like sac.
- It lies in the lumbosacral region.
- It result in neurological deficits, but they are usually not associated with mental retardation.
- It may be of following 3 types:

a. Spina bifida with *meningocoele*- When only fluid ûlled meninges protrude through the defect.

b. Spina bifida with *meningomyelocoele*- When neural tissue is included in the sac.

c. Spina bifida with *myeloschisis* or *rachischisis*- When occasionally the neural folds do not elevate but remain as a flattened mass of neural tissue.

Spina bifida with meningomyelocoele is a more common and a more severe defect than spina bifida with meningocoele. This neural tube defect can occur anywhere in vertebral column, but lumbosacral region gets commonly involved [1]. Child with meningomyelocoele presents with a cystic swelling at lumbosacral region covered with or without weeping skin, sometimes with ruptured membranes. Spinal cord will be on open platform covered with meninges which is liable to infections [8]. When the membrane ruptures the ultimate outcome will be meningitis secondary to infections. They also complain decreased motility of lower legs and dribbling of urine due to dysfunction of cauda equina roots or conus medullaris contained in the sac [9]. Our patient had lower limb hypotonia. About 90% of patients of meningomyelocoele will present with hydrocephalus. Hydrocephalus is a Greek word meaning hydor "water" and kephale "head" due to excess cerebrospinal fluid (CSF) accumulation in the head. As spinal cord gets tethered to the vertebral column, it gets elongated. This tethering pulls the cerebellum downward & thus occluding the foramen magnum. Hence there is cutting off the ûow of cerebrospinal ûuid resulting in the development of hydrocephalus. Hydrocephalus ultimately causes bulging fontanelle, enlarged scalp veins, macrocrania, suture diastasis, and positive Macewen (ie, cracked pot) sign and sunset sign. Our patients had hydrocephalus with wide open and bulging fontanelle and positive sunset sign. Meningomyelocoele patients with hydrocephalus often presents with Arnold Chiari II Malformation, an abnormal downward herniation of the cerebellum and brain stem through foramen magnum. If untreated they develop headaches, blurred vision, decline in intellectual performance, and gradual drowsiness, which, lead to coma and death due to respiratory arrest. There are some prenatal screening tests that are used to detect congenital anomalies. For example quadraple test done in second trimester detects meningomyelocoele, Down's syndrome and other congenital malformations. Patients carrying baby with with spina bifida will have increased AFP levels in blood higher than the normal. Currently, in-utero USG scanning can detect hydrocephalus at an early intrauterine life. This permits good identification of any ventricular dilatation that indicates active hydrocephalus. In a suspected case of hydrocephalus CT scan confirms the diagnosis. In selected cases amniocentesis can be performed [10]. Regular ANC visits in each trimester and folic acid & vitamins supplements taken in periconceptional period have shown to reduce the incidence of neural tube defect by 70% [1]. After the baby is born with meningomyelocoele, a surgical closure is indicated under full antibiotic control because of risk of meningitis. Hydrocephalus is managed by ventriculoperitoneal shunt in which excess CSF is continuously drained in peritoneal cavity to normalize the pressure. Though with successful treatment a close follow up of these patients is mandatory [10].

Conclusion

Spina bifida with meningomyelocoele with hydrocephalus is rare congenital anomaly with incidence of 1 in 1000 live births that is associated with fetal mortality and morbidity. Prenatal screening for congenital anomalies and nutritional supplement of folic acid can lower the incidence of neural tube defects.

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