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A Multimodal Treatment Approach on Rare Case of Meningomyelocele

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Meningomyelocele, one of the neural tube defect (NTD) variants, is a condition in which meninges and neural tissue protrudes through a defect in vertebral arch and forms a cyst-like sac with an incidence of 0.5–11/1000 live births. Hydrocephalus is a major problem for majority of patients with meningomyelocele due to the abnormal accumulation of cerebrospinal fluid (CSF) in the ventricles of the brain. Meticulous preoperative evaluation with planning of the anesthetic technique, especially in regard to managing the airway, plays a crucial role in successful surgical management of these cases. Here, we report a case of lumbosacral meningomyelocele with hydrocephalus underwent a multimodal treatment approach for providing a comprehensive plan of care.

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1. INTRODUCTION

Meningomyelocele or spina bifida is a congenital neural tube defect that results in the malformation of the spine and a potential dysplastic spinal cord [1]. The severity of defect ranges from spina bifida occulta upwards to anencephaly [2]. Recent estimates suggest that spina bifida without anencephaly is the most prevalent non-chromosomal central nervous system (CNS) defect with an occurrence rate of 3.73 per 10,000 births in the United States and ranks 7th among other non-CNS birth defects based on the data by Centers for disease control and prevention (CDC) [3][4].

2. CASE PRESENTATION

A seven month old baby girl came to the pediatrics out patient department (OPD) with a chief complaint of congenital lump of mass in the lumbosacral region. The approach of the baby's parent were for the surgical excision of a cystic swelling in lumbosacral area present since birth.

The gestational history of the mother was presenting a full term normal vaginal delivery with no complication but the baby presenting a defect in lumbosacral region in midline. Mother gave history of gradually increasing swelling since birth to present size of 2 cm in diameter with leaking and bleeding from the site. Mother also noticed increase in the size of head since 4th month of chronological age. There is also a history of admission in hospital after birth due to low birth weight (2 kg) and bleeding from the lumbosacral swelling site.

Baby was afebrile to touch with a heart rate (HR) of 132 beats per minute, respiratory rate (RR) was 31 breaths per minute and oxygen saturation was 98%.

The systemic examination included respiratory examination showing vesicular breathing sound which were normal in all the lung fields. The cardiovascular system examination noted the first and second heart sound were heard with no murmurs. The baby was conscious, cooperative, active and well oriented to time, place and person as per the findings of central nervous system examination.

The local examination of spine was showing cystic swelling which was soft in nature on the lumbosacral(LS) region and skin over the swelling was intact, stretched and thin which was on the verge to burst as shown in Fig. 1.



Fig. 1. The cystic swollen mass on the lumbosacral region

2.1 Radiological Investigation

The radiological findings were showing an evidence of a bony defect in lumbosacral region with protrusion of meninges and cerebrospinal fluid (CSF) with spinal cord along with the sac covered by skin as shown in figure 2. Also, there was an evidence of tethered cord and syrinx formation from T12-L4 level as shown in figure 3. The figure 4 is showing the gross hydrocephalus with herniation of peg like cerebellar tonsils.

2.2 Management

The initial surgical intervention was closure of open duct generally done on first day of life. The hydrocephalus is very common and can occur at any time but most frequently within few months of life. The ventricular dilation may process the change in head circumference or sings of increased intracranial pressure. The treatment of ventriculomegaly was mild limited to clinical observations. In advanced cases surgical placement of a ventricular shunt or endoscopic 3rd ventriculomegaly is done. The orthopedic complications are common in myelomeningocele such as scoliosis, lordosis and kyphosis and the management includes TLSO (thoracic-lumbar-sacral orthotics) and proper seating habits. The surgically implanted growing rods to support developing spine is proved to be a boon in the management of such cases.

The medical management included the anticholinergic to suppress the overactivity of detrusor for example oxybutynin chloride. The Tricyclic antidepressant such as hydrochloride or imipramine plays an important role. The alphaadrenergic antagonist are used as it decreases the bladder outlet resistance and increases urinary flow rate and improve the bladder emptying.



Fig. 2. Evidence of a bony defect in lumbosacral region with protrusion of meninges and cerebrospinal fluid (CSF) with spinal cord



Fig. 3. Evidence of tethered cord and syrinx formation from T12-L4 level

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Fig. 4. Gross hydrocephalus with herniation of peg like cerebellar tonsils

The Physiotherapy care included the close monitoring of the developmental milestones of the baby with maintain the normalcy of the same. The physiotherapeutic approach was mainly to prevent any expected secondary complication of the condition. The maintenance of the lung compliance and activeness of the baby were mandated throughout the physiotherapy treatment.

Also, the immediate care includes putting the patient in face down position, curtain the area which is affected with gauze piece (sterile) which is dipped in normal saline, maintenance of fluid balance and monitoring of associated defects.

The long term line of treatment includes the use of braces and catheters with close monitoring of the diet rich in high fibers and the antibiotics can be used to prevent or treat infections like urinary tract infection (UTI).

3. DISCUSSION

The pathophysiology following the meningomyelocele includes estimated failure in the fusion of neurulation in the primary stage during the day 21-26 of gestation approximately [5]. The failure in fusion are hugely manipulated by the endogenous factors as well as exogenous factors [6]. The deficiency of folate is found in 70% of cases of meningomyelocele interfering in the fusion and resulting in the defect in the placode architecture [7]. The exogenous factors vary from the gestational course to the environmental toxins that may hinder in the fusion of neurulation [8].

The presentation of hydrocephalus along with the cystic swollen mass has reduced with more stringent criteria placed on shunt but still being presented by 80% of children suffering from meningomyelocele [9]. The malformation of chiari 2 as a result of defective CSF flow in hydrocephalus may emerge from stenosis of aqueduct, obstruction of fourth ventricular outlet or subarachnoid space obliteration following posterior fossa crowding or tentorial hiatus narrowing [10].

4. CONCLUSION

The multimodal approach of treatment has resulted in the accelerated step-wise recovery of the baby suffering from the meningomyelocele. The neural tube defect as presented and treated has given an effective line of treatment along with the management of hydrocephalus for an uninterrupted and improved quality of life of the baby.

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INFORMED CONSENT

Written & Oral informed consent was obtained from the participant included in the study.

ETHICAL APPROVALS

We conducted our research after obtaining proper IEC approval.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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