

Case Report

A newborn with thoracic and lumbosacral meningocele without neurological deficit

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Thoracic meningocele and myelomeningocele are rare neural tube defects unlike lumbosacral neural tube defects; sometimes there no neurologic deficits and thus the subtle features of cervical cord tethering may be overlooked on imaging. The presence of neural tube defects at multiple (two or more) sites along the vertebral axis is a very rare event occurring in less than 1%

of cases. Up to our best knowledge less than 10 cases have been reported in the literature. We report a case of multiple neural tube defects (thoracic and lumbar) in same patient with no any neurological deficit found. (Rawal Med J 201;41:382-384)

Key words: Meningocele, myelomeningocele, neural tube defects.

INTRODUCTION

Congenital fusion defects of neural tube and associated tissues are classified as spinal dysraphism. Neural tube defects vary in severity and affect the tissues overlying the spinal cord; meninges, vertebral arches, muscle and skin. Amongst the congenital defects of CNS, spina bifida cystic (myelomeningocele) is the most common.¹⁻³ The incidence for spinal dysraphism has been reported to be 1 in 800 live births.¹⁰ A sac like covering is filled with CSF, and may contain spinal cord and nerve roots called myelomeningocele or may not contain spinal cord and nerve roots and called meningocele. Associated findings are those of Arnold-Chiari malformation, which include hindbrain herniation into upper cervical spinal canal, small posterior fossa, dysgenesis or agenesis of corpus callosum, mucosal migration disorder of varying degrees and hydrocephalus.^{4,5} Previous family history of affected child increased risk for neural tube defects; maternal folic acid deficiency is an environmental factor that strongly associated with neural tube defects.^{1,5} Demographically, North India is a high incidence area for spinal dysraphism; Antenatal diagnosis can be done with ultrasonography and fluid Alfa-fetoprotein in amniotic fluid.

CASE PRESENTATION

A three weeks old male child, born by cesarean

section to 30 years old mother gravida 6, para 5, at gestational age of 37 weeks with Apgar score 6/10 after suction and bagging 8/10, weighed 2.3 kg, had head circumference of 33 cm and length 45 cm. No antenatal ultrasound was done. Examination revealed two neural tube defects, one meningocele mid thoracic and the other myelomeningocele at the lumbosacral area (Fig.). There was no leakage of CSF. The sac in thoracic region measured 35*25 mm with bone part from spinous process and lamina, and in the lumbosacral region 55*45 mm. No neuromuscular or sphincter involvement but there was dilated ventricles without bulging in the anterior fontanel or sings of hydrocephalus was observed.

Fig. Pre operative appearance.



Surgical repair of myelomeningocele was performed. In a follow up period of 4 months, the patient had communicated hydrocephalous and VP shunt was inserted.

DISCUSSION

Most congenital anomalies of the spinal cord vary from a minor spina bifida occulta to a more clinically significant spina bifida cystica, involving spinal cord and meninges in addition to the vertebral arches.^{6,7} Cervical and thoracic regions are the least common sites, and lumbar and lumbosacral regions are the most common sites for neural tube defect. Neonates with a meningocele usually have normal examination findings and a covered dural sac. They usually do not have associated neurologic malformations such as Chiari malformation type 2 or hydrocephalus.^{8,9}

Myelomeningocele is a condition in which the spinal cord and nerve roots herniate into a sac comprising the meninges. The spinal cord often ends up in this sac in which it is splayed open, exposing the central canal. The splayed open neural structure is called the neural placode. Myelomeningoceles have a higher incidence of associated intestinal, cardiac, and esophageal malformations, as well as renal and urogenital anomalies.^{4,10} Most malformations, especially those such as neural tube defects, occur early in embryogenesis and are likely the result of aberrant expression of a yet undefined developmental gene or family of genes.

The nervous system develops in a precise temporal embryologic sequence; therefore, an interruption of one part of the developmental sequence often affects remaining development. Double myelomeningocele is a very rare event. Bertran and Wilson described a double myelomeningocele similar to our case at the thoracic and lumbosacral levels in a patient who presented at about two weeks after birth with infection and CSF leak.² This patient also had hydrocephalus and Arnold-Chiari malformation with lower extremity neurological deficits.

In a series by Erashin et al⁵ most children with myelomeningocele had Para paresis. Another case was reported which was present in thoracic and

lumbar region.¹⁰ The first case described is rarer as very few cases of meningocele have been reported in the cervical region. Also, during surgical repair, a diastomyelia due to fibrous spur was detected, which was excised. The second case, like most of other cases, was in lumbar and lumbosacral region. Associated anomaly of club feet was also present and post operatively, the patient developed hydrocephalus as a complication of surgery.

In summary, the most important aspects in evaluating the outcome of neonates with myelomeningocele are continence, hydrocephalus, orthopedic problems, intellect, ambulation, and social aspects of life (employment and independent living status).

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