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Meningomylocele – A case report

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Abstract

Meningomylocele is the most common abnormality of the neural tube. Cervical meningomylocele is less common among them. The embryological basis and clinical features are different from thoracolumbar and lumbosacral, which are more commonly, occur. Meningomylocele is protrusion meninges with accompanying nervous tissue through a defect in the posterior vertebral arches. In this study, we have reported a case of cervicalmeningomylocele.

Keywords: Meningomylocele, Neural tube, Cervical meningomylocele

Introduction

Neural tube defects are the commonest malformations leading to fetal wastage. Commonest malformations of central nervous system are neural tube defects, which include Anencephaly, Encephalocele, Spina bifida – Meningocele and Meningomylocele. The incidence of neural tube disease varies from country to country and region to region within country. The highest incidence was reported from Ireland and lowest from South America. Meningomylocele is protrusion of meninges with accompanying nervous tissue through a defect from posterior vertebral arches and associated with neurological deficit.Usually evident as a fluid filled protrusion covered by skin or membrane in the midline. The clinical characteristics are readily observable, palpable, protuberant and fluctuant lesion. Varying degree of paresis of the legs and sphincter dysfunction are the major clinical manifestation. Congenital dislocation of hip or deformities of foot may occur. Severe sensory loss and accompanying tropic ulcers may prove vexing (Habibi *et al.,* 2006; Jaeger *et al.,* 1997).

The normal sites of meningomyelocele are lumbosacral, thoracolumbar and cervical among these lumbosacral involvement is more common this type may produce lower motor neuron signs due to abnormalities and disruption of the conus medullaries. Thoracic defects are more complex and frequently associated with serious complications. The conditions with a meningomylocele in the upper thoracic and cervical region usually have a minimal deficit. The embryological cause for meningomylocele is failure of closure of neural tube during third week of gestation leads to the constellation of defects. The open neural tube is continuous with the surface of the skin for this reason infants with meningomylocele are at risk for bacterial meningitis due to the spinal defect. Leak of cerebrospinal fluid is commonly observed. The major indication for early operative repair within 48hr of delivery is prevention of infection. Although protection of the exposed

neural tissue from trauma and drying caused by meningomylocele is fixed and rarely improves following repair. Deterioration, however, can occur (May *et al.,* 1992; Pang & Dias, 1993; Pang *et al.,* 1992).

The associated defects with meningomylocele are hydrocephalus a frequently associated defect is the result of the Arnold chiari malformations they may be associated aqueductal stenosis. Hydrocephalus present in 73% of cases with meningomylocele. The meningomylocele is most common with thoracolumbar lesions such as anomalous rib formation; fused or bifid rib is very common. Kyphotic angulations of thoraco lumbar spine one or more hemi vertebral causing scoliosis can also be associated. True developmental malformations of the urinary tract are found these includes contra lateral fusion, dysplastic, duplex and polycystic kidney. The genetic cause for meningomylocele transmitted as an autosomal recessive trait, although recurrence risk statistics suggests of polygenic or environmental etiology (Salomao *et al.,* 2006; Steinbok & Cochrane, 1991).

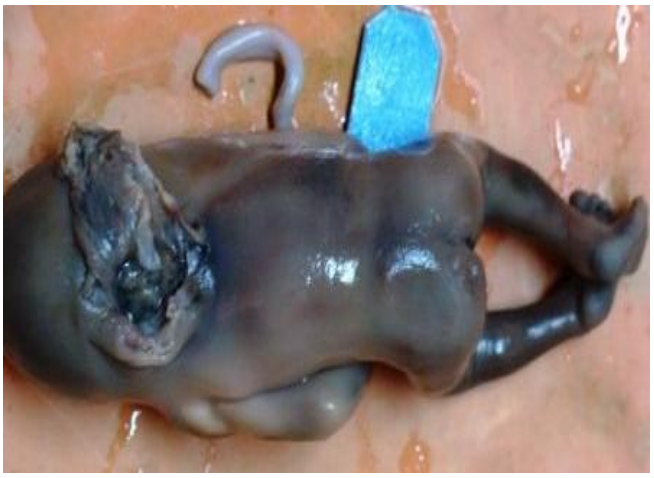
Case report

- We have received an aborted fetus with cervical meningomylocele from OBG department, JJMMC, Davangere.
- The history of mother was not available.
- The fetus weight was 2300gm.
- After examination we have found a ruptured large mass sac in the cervical region.
- In the sac protrusion of the meningeal layers with extent of neural involvement through a defect in the boney encashment of the vertebral column that is Spina Bifida.
- Spina Bifida is a developmental anomaly characterized by defective closer of vertebral arches/body.
- Hydrocephalus observed (Fig-1).

Discussion

Meningomylocele is a complex congenital spinal anomaly that causes varying degrees of spinal cord malformation. It is commonly referred to as Spina Bifida and is a developmental anomaly characterized by defective closure of the bony encashment of the spinal cord and meninges may or may not protrude. Patients with meningomylocele present with a spectrum of impairment but the primary functional deficits are lower limb paralysis, sensory loss bladder, bowel dysfunction and cognitive dysfunction. Cervical meningomylocele is a relatively uncommon group of spinal dysmorphism with a very low incidence as compared to the more commonly occurring thoracolumbar and lumbosacral type (Vogter *et al.,* 1987; Steinbok & Cochrane, 1991).

Fig.1. Showing the cervicomeningomylocele



Cervical meningomylocele and thoracolumbar meningomylocele have more unique features but it differs from thoracolumbar meningomylocele are follows associated Chairi II malformations is less common, full thickness skin covers, intact neurological functions and content is not a misplaced neural placode. The limited dorsal myloschisis theory of pathogenesis have been explained better all above features. In all neural tube defects the occurrence of cervical meningomylocele is less than 5% and with less motor deficits, urinary voiding difficulties and deformities of urinary system associated than thoracolumbar and lumbosacral meningomylocele which are classified as lower neural defects (Pang *et al.,*1992). In open neural tube defects the more commonly reported 44-62% cases of cervical

meningomylocele associated with Chairi II malformation. Cervical meningomylocele is also associated with hydrocephalus but compare to other meningomylocele it is less. Cervical meningomylocele are two types: one is more common that is fibro neurovascular stalk extending from the cervical part of spinal cord into the living of the sac and other one is less common that splits into two hemi cords separated by a midline fibrous septum (Pang & Dias, 1987; Eller & Bernstein, 1987; Kulakarni *et al.,* 1989).

The repair of meningomylocele is surgical treatment only that is simple and has evolved over time from just simple extradural excision to a more comprehensive surgical excision consisting of resection of the lesion, intradural exploration to detach the cord if fixed by a fibrous band and management of associated hydrocephalus with the recognition that children operated by only extradural exploration developed neurological deterioration on follow up (Scott *et al.,* 1994). Neurological outcome is generally good immediately but may sometime need secondary surgical exploration after retethering or failure to address the associated lesion during primary surgery (Rodney Harris & Read, 1981).The diagnosis of meningomylocele is usually diagnosed prenatally by ultrasound during the second trimester. Positive screening for maternal serum alpha-fetoprotein may also prompt a fetal ultrasound. Postnatal diagnosis usually by MRI investigations. Our case report may help to update of knowledge about cervical meningomylocele.

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