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Case Report

Cervical meningocele with tethered cord

Rugmini Kamalammal*, Siddaraju Malaveli Lingappa, Palmurthy Rajalu

Department of Pediatrics, Adichuchanagiri Institute of Medical Sciences, Mandya District, Karnataka, India

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***Correspondence:**
Dr. Rugmini Kamalammal,
E-mail: rugminirao@yahoo.com

ABSTRACT

Cervical meningocele is a spinal dysraphism which is occurs rarely. It can present with associated tethered cord, if so here is a of the child developing neurological deficit later on in life. Here we are presenting a case of new-born with tethered cord and no neurological deficit at present .The treatment is aimed at prevention of development of the neurological deficit.

Keywords: Cervical meningocele, Tethered cord, Excision

INTRODUCTION

Incidence of cervical meningomyelocele and meningocele is about 1-5 % of neural tube defects. Spinal dysraphisms at the level of cervical region is rare as seen in a reported series.[1,2] Same author classified the lesion into 3 types - type I - a fibrovascular or neuroglial tissue protruding from posterior surface of spinal cord attach to the sac wall, type II - an ependymal-lined cyst that herniates inside of a meningocele, representing a hydromyelic canal in connection with an outer cyst and type- III true meningoceles in which meningeal tissue herniates through the defect and the sac contains only CSF.[2]

Type 1 is commonly seen and can be associated with tethered cord. Other associated anomalies include hydrocephalus, Chiari malformations, hydromyelia, diastematomyelia, thickened filum terminal and Klippel-Feil syndrome.

CASE REPORT

A term new born baby was delivered by caesarean section to a 26-year old primigravida in our institution out of non consangiuous marriage. Her perinatal history was unremarkable. She had regular adequate folic acid, iron and calcium supplementation during the pregnancy. Baby had birth weight, Length and Head circumference of 2.8 kg, 48 cm, 34.5 cm respectively (Within normal limits for gestational age). Baby cried immediately after birth. On examination, a lemon shaped round mass was noticed on the posterior part of the neck. The sac was round, fluctuant, and had a wide, sessile base covered with full thickness skin except the apex of the sac which was reddish blue in colour and was covered with only a thin membrane (Figure 1).

There was no associated neurological deficit. USG abdomen was normal. MRI of the head and spine showed a well-defined fluid filled cyst with few septations from the subcutaneous tissue to skin at posterior midline of neck, protrusion measuring 4.2×3.7×2.9cm. Posterior tented bulge of cervical spinal cord is seen at C4/C5 level, posterior tent of dorsal dural sac is also noted at the same level. A low signal connection between the posterior bulge of the cord and the dural sac extending to cyst wall, representing tethered cord was also seen. Rest of the spinal cord and brain appeared normal. There was no sign of cerebrospinal fluid leakage. Unfused spinous process of multiple vertebrae noted (Figure 2).

DISCUSSION

Our case has a fibrovascular tissue protruding from posterior surface of the cord and attached to the sac wall,

classifying it as Type 1.There is a tethered cord which is prone for development of neurological deficits later on in life.[3]

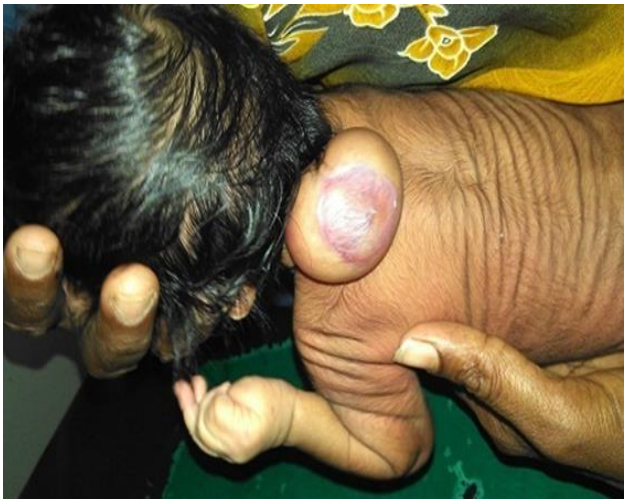

Figure 1: A case of meningocele.

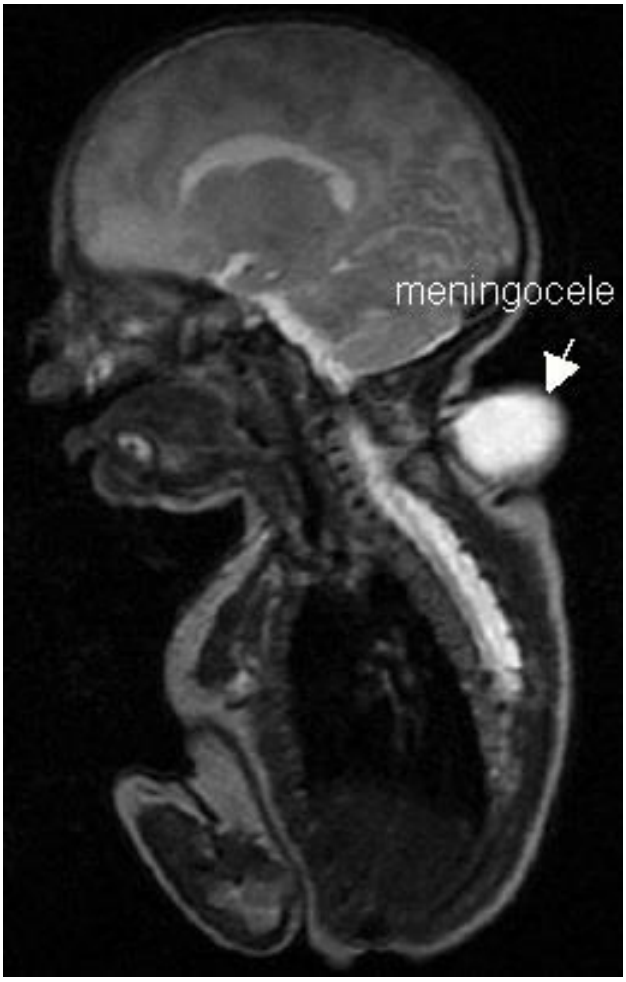

Figure 2: Unfused spinous process of multiple vertebrae.

Neural tube defects are multifactorial in origin. Studies have shown that periconceptional folic acid intake can prevent this anomaly. Lack of folic acid causes increase in Homocysteine levels in the serum which may be a reason for occurrence of this anomaly. But in our case the mother was on folic acid in the 1st trimester but not during conception.[4] If one parent is affected the chance of sibling having this anomaly is 1in 30.[5]

The chance of high degree of movements in cervical region stretches the cord in the tethered region and can cause further neurological defects.[4] over the years with motor function in the upper extremities being primarily affected.

Diagnosis is done by MRI which will give the type of anomaly and also associated anomalies in detail. Ante natal diagnosis is by maternal serum alpha fetoprotein (MSAFP), amniotic fluid alpha fetoprotein and amniotic fluid acetyl cholinesterase. Screening by elevated MSAFP measured at 16-18 weeks of gestation can detect 56-91% of affected fetuses.[5,6]

Ultrasound is a good tool for screening and has a sensitivity of 79-96% and specificity of 90-100%.[5.7]

Most posterior meningoceles contain aberrant nerve roots adherent to the inner wall, occasional ganglion cells or even glial nodule that may represent a diverticulum of the central canal of the spinal cord. It could be either due to a taut fibro-neurovascular stalk that extends from the dorsal column of the cord to the dorsal dural of the sac namely dorsal myeloschisis or it occurs as a cervical myelomeningocele that contains a type II median fibrous septum split cord malformation, a dorsal band of connective tissue, nodules within the sac, an ependymal lined cavity.[8,9] In this situation, a between the two hemi cords tethers them to the dorsal dura.

Surgical excision of the meningocele as well intradural exploration to untether the spinal cord is extremely necessary to prevent delayed neurological deterioration, prevention of neurological deficit by releasing tethered elements and cosmetic reasons. Furthermore, hydrocephalus may coexist and careful management, with VP shunt, is mandatory.

CONCLUSION

Cervical meningocele is a very rare malformation which to some extent can be prevented by preconceptional folic acid supplementation. Even though there is no neurological deficit at present it is ideal to do surgery at the earliest to prevent development of such neurological complications.

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