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Spina Bifida Cystica – Meningocele

KEYWORDS: neural tube defects, spina bifida, meningocele, meningo myelocele, rachischisis.

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ABSTRACT *Back ground: most of the congeniltal anomalies of spinal cord are due to non closure of neural tube in 3rd wk of gestation. Most of the defects affect the tissues overlying the spinal cord that is meningeal coverings , vertebral arches , muscles and skin. spinabifida is referred to defective vertebral arches . when the defect is severe it leads to protrusion of meninges called meningocele. Protrusion of parenchyma of spinalcord along with meninges leads to formation of meningomyelocele.*

Method: The present case is one amongst the neural tube defects presented in the literature. 50 dead foetuses were brought from government maternity hospital and various other private hospitals for post graduate dissertation works and all the dead fetuses were infused with formalin solution for preservation of the fetuses. The Crown Rump length of fetuses were measured and the gestational age was calculated.

Result: A 28 weeks old foetus presented certain abnormalities in the mid-line at the level of lumbar region in the form of a sac. A further examination in sac revealed the protrusion of meninges only with no incorporation of nervous tissue in the walls of the sac. The entire ssac is covered by very thin skin. There is no evidence of associated defective calvaria that is absence of cranio lacunia. There is no evidence of myeloschisis

Coclusion: it is a case of Spina Bifida-Cystica.To provide information regarding the use of folic acid in combination with a multivitamin supplement for the prevention of neural tube defects and other congenital anomalies, so that physicians, midwives, nurses, and other health care workers can assist in the education of women in the pre-conception phase of their health care.

INTRODUCTION:

Most of the congenital anomalies of spinal cord are due to non closure of neural tube in 3rd wk of gestation. Most of the defects affect the tissues overlying the spinal cord that is meningeal coverings , vertebral arches , muscles and skin. Spinabiufida us referred ti defective vertebral arches. When the defect is severe it may leads to protrusion of meninges called meningocele . protrusion of parenchyma of spinal cord along with meninges leads to formation of meningomyelocele. Sometimes a small sinus may be present in the sacral region representing the last piece of separation between surface ectoderm and neural tube . the present case is one amongst the neural tube defects presented in the literature.

METHOD:

50 dead foetuses were brought from government maternity hospital and various other private hospitals for post graduate dissertation works and all the dead fetuses were infused with formalin solution for preservation of the fetuses. The Crown Rump length of fetuses were measured and the gestational age was calculated. A 28 weeks old foetus presented certain abnormalities in the mid-line at the level of lumbar region in the form of a sac. By using the dissection instruments while doing the dissection we observed the defects and following observations were noted.

OBSERVATIONS:

An external examination of the foetus revealed a sac like structure in the lumbar region. the sac is protudilng through the defectilve vertebral arches at the lumbar region . A further examination in sac revealed the protrusion of meninges only with no incorporation of nervous tissue in the walls of the sac. The entire ssac is covered by very thin skin. There is no evidence of associated defective calvaria, that is absence of cranio lacunia. There is no evidence of myeloschisis . When spina bifida cystica is manifested severly it results in partial absence of brain called meroanencephaly . this condition shows varying degree of neurological deficit depending on the position and extent of lesion. Sometimes only an area of anaesthesia is manifested in all cases of meningomyelocele , sphincters are affected results in loss of sensation in the region that impinges on the saddle during riding.cases of spinabifida donot always results in mortality. But here in this case no such abnormality is found.

DISCUSSION AND CONCLUSION:

spina bifida cystica with meningocele occurs 1in 1000births. In the british isles the incidence vaies from 4.2 per 1000 new born infants (south wales) and 1.5 per 1000 insouth eastern England.

Suspicion of spinabifida cystica and meroanencephaly in utero can be considered when high levels of alpha feto protein in amniotic fluid are found. An ultrasound examination of the foetal vertebral column at 2 to 12 weeks gestation will be very helpful to diagnose spinabifida cystica[1].

Maternal serum alpha foeto protein (MSAFP) determnation will be more useful in all cases of neural tube defects in the foetus seem to be there is a scope for escape of more AFP into amniotic fluid[2].

To gain utility of MSAFP test the gestational age must

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be known with certainty. This is because the amount of MSAFP increases with gestational age(as the foetus and the amount of AFP produced increase in size).Also the race of the mother and presence of gestational diabetes are important to know because MSFAP Can be affected by these factors.

The MSAFP is typically reported as multiples of the means (MOM). The greater the MOM the more likely the defect is present . MSAFPhas greatest sensitivity between 16&18 weeks of gestation but can still be useful between 15&22 weeks of gestation. The most common cause of elevated MSAFP wrong estimation of gestational age of the foetus.

Using a combination of MSAFP screening and ultra sonography almost all cases of spinabifida can be distinguished from other foetal defects. If acetyl choline esterase is elevated along with MSAFP a neural tube defect is most likely[3,4].

The neural tube defect resulting in spinabifida is due to genetic polymorphism a result of mutation in methylene tetrahydrofolate reductase gene.Folate is co-factor for this enzyme which play an important role in homocysteine metabolism Any mutation at the position of nucleotide 677 and 1298 in methylene tetrahydrate gene leads to abnormal gene code associated with elevated maternal homocysteine concentration and this results in increased risk for NTDs in fetuses.

The recommended strategy to prevent recurrence of a congenital anomaly (anencephaly, myelomeningocele, meningocele, oral facial cleft, structural heart disease, limb defect, urinary tract anomaly, hydrocephalus) that has been reported to have a decreased incidence following preconception / first trimester folic acid with multi vitamin supplementation[5,6,7]. The recommended strategy(ies) for primary prevention or to decrease the incidence of fetal congenital anomalies will include a number of options or treatment approaches depending on patient age, ethnicity, compliance, and genetic congenital anomaly risk status. For Patients with no personal health risks, planned pregnancy, and good compliance require a good diet of folate-rich foods and daily supplementation with a multivitamin with folic acid (0.4-1.0 mg) for at least two to three months before conception and throughout pregnancy and the postpartum period (4-6 weeks and as long as breastfeeding continues). Patients with health risks, including epilepsy, insulin dependent diabetes, obesity with BMI >35 kg/m2, family history of neural tube defect, belonging to a high-risk ethnic group (e.g., Sikh) require increased dietary intake of folate-rich foods and daily supplementation, with multivitamins with 5 mg folic acid, beginning at least three months before conception and continuing until 10 to 12 weeks post conception.

Minimally invasive fetoscopic surgery for spina bifida has been developed to improve the postnatal neurological function of affected fetuses and to achieve a reduced maternal trauma compared to open fetal surgery, regardless of placental position[8].

Open fetal surgery for spina bifida seems to show lower rates of procedure-related complications compared to endoscopic surgery, however high rates of hysterotomy scar complications are still found after open surgery[9].

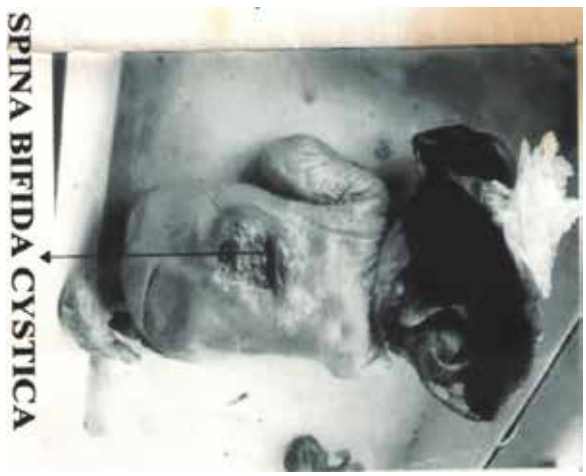

Figure 1: Showing incomplete fusion vertebral arch with protrusion ofmeninges

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