MEGA CISTERNA MAGNA DIAGNOSTIC DILEMMA

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ABSTRACT

Objective: To determine the frequency of isolated mega cisterna magna (MCM) in Saudi population, compare the accuracy of fetal ultrasound with postnatal neuro-imaging, report the associated central nervous system (CNS) anomalies on postnatal neuro-imaging.

Study Design: Retrospective study.

Place and Duration of Study: King Fahad Armed Forces Hospital Jeddah Saudi Arabia from Jan 2007 to July 2011.

Material and Methods: This was a retrospective study conducted at King Fahad Arrmed Forces Hospital Jeddah, Saudi Arabia. We included all reported cases of fetal mega cisterna magna from January 2007 till July 2011. Ante-natal and postnatal records and neuro-imaging studies of all the cases were reviewed and documented on the database. Neurological assessment was done by Pediatric Neurology Consultant at an average age of 8 months of life.

Results: MCM was diagnosed in 26 (0.12%) on fetal scans (15 male and 11 females). Post natal scans were available for 20/26 (77%) and 6/26 (23%) cases were missing. Of the 20 post natal scans, 12 (60%) scans were normal and 8 (40%) scans were abnormal. The following abnormalities were detected: 3 (37.5%) cerebellar hypoplasia, 2 (25%) partial dysgenesis of corpus callosum, 2 (25%) cases were confirmed as MCM and 1 (12.5%) arachnoid cyst. Neuro-developmental assessment was reported as normal in 50% of the cases with associated CNS anomalies.

Conclusion: The association of MCM with major CNS anomalies is significant in this study population. Post natal neuro-imaging confirmation is required for all fetuses with mega cisterna magna on fetal sonography.

Keywords: Head ultrasound, Mega cisterna magna, MRI brain.

INTRODUCTION

A mega-cisterna magna (MCM) is defined as a dilated cisterna magna whereas fourth ventricle, cerebellar hemispheres and vermis are normal[1]. The cistern space is present between the cerebellum, the dorsal surface of the medulla oblongata and the dura lining of occiput. Cerebro spinal fluid (CSF) drains to cisterna magna from the fourth ventricle through the lateral apertures and median aperture.

MCM is reported in 1% of normal population postnatally[2]. Clinical significance of isolated MCM is not clear and most of the studies so far reported a good prognosis[3]. An enlarged cisterna magna has been associated with chromosomal abnormalities such as trisomy 13 and 18[4]. In addition, it has also been reported in cases of trisomy 21[5]. The developmental outcome of most of the children with isolated MCM were normal[6].

The diagnostic modalities of posterior fossa malformations include cranial ultrasound and brain MRI both in ante-natal and postnatal set up. If the vertical distance between the vermis and the inner border of the skull is more than 10 mm on ultrasound, it indicates a mega cistern magna. It still remains challenging to diagnose posterior fossa dysgenesis in ante-natal scans despite the availability of modern neuro-imaging techniques because of high probability of false positive and negative diagnoses[7]. When stratified by gender, male fetuses characteristically have higher frequency of isolated MCM compared to female fetuses and the mean measurements of the cisterna magna appear to be larger for males[8]. MCM can

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be a part of complex central nervous system anomalies, which alters the prognosis and long term outcome[9].

MATERIAL AND METHODS

It is a retrospective study conducted at a tertiary care teaching hospital King Fahad Armed Forces Hospital, Jeddah, Saudi Arabia. All babies born with fetal isolated MCM between January 2007 and July 2011 were included. Fetal anomaly scan was done at 20 weeks of gestation by sonographer, and if anomalies were suspected, the case were referred to obstetrics and gynecology sonologist for confirmation and follow up studies. Postnatal head ultrasound was performed on day 1 of life to confirm antenatal diagnosis. If postnatal head ultrasound was normal, the cases were discharged without follow up, if found abnormal, MRI brain was done. If MRI brain was abnormal, follow up appointment was given for neurological assessment. We excluded the cases with multiple congenital malformations and intra uterine fetal deaths. A total of 26 fetuses with isolated MCM were selected by purposive non probability sampling. Ante-natal and postnatal records with imaging studies of all the cases were reviewed and documented on the database. Neurological assessment was done by Pediatric Neurology Consultant at an average age of 8 months and if abnormal further neurological follow up assessment was advised.

Data were entered on Statistical Package for Social Sciences 20 (SPSS inc., Chicago, IL,

Table-1: Postnatal abnormal neuro-imaging of eight cases with fetal isolated MCM.

Fetal sonography	Postnatal Head ultrasound	Postnatal MRI
Isolated MCM	Slightly prominent cisterna magna	Mild prominence of cisterna magna with some degree of cerebellar hypoplasia
Isolated MCM	Slightly prominent cisterna magna	Mild prominence of cisterna magna with some degree of cerebellar hypoplasia
Isolated MCM	Slightly prominent cisterna magna	Posterior fossa suggestive of arachnoid cyst Mild prominence of
Isolated MCM	Slightly prominent cisterna magna	Mild prominence of cisterna magna with some degree of cerebellar hypoplasia
Isolated MCM	Slightly prominent cisterna magna	Mega cisterna magna with wide CSF spaces around cerebellum (Isolated MCM)
Isolated MCM	Slightly prominent cisterna magna	Mega cisterna magna with wide CSF spaces around cerebellum (Isolated MCM)
Isolated MCM	Partial dysgenesis of corpus callosum	Not done
Isolated MCM	Partial dysgenesis of corpus callosum	Partial dysgenesis of corpus callosum

MCM (Mega cisterna megna), CSF (cerebrospinal fluid).

USA) and analyzed. Simple descriptive statistics were used like mean and standard deviation (SD) calculated for quantitative variable like age. Frequencies and percentage were used for qualitative variables like gender, detection of MCM on antenatal and postnatal scans. This study was conducted according to principles of Helsinki declaration.

RESULTS

Total number of deliveries were 25443 and 4100 (16%) were un-booked during the study period. Isolated MCM was detected in 26 fetuses (0.12%) on antenatal fetal scan. All of them were born alive (1/978 births) 15 (58%) were males and 11 (42%) were females. Postnatal head scans were available for 20/26 (77%) in the form of head ultrasound and MRI brain and data for 6 cases could not be available. Two delivered outside and 4 cases were discharged without postnatal scan with appointment but lost follow up. A total of 9 (45%) postnatal MRI brain were available for evaluation of which 2 MRI were without head ultrasound. MRI brain of these two cases were normal. Postnatal neuro-imaging studies were normal in 12 cases (60%) and 8 (40%) were abnormal. False positive antenatal scans were (60%). All the neonates who had abnormal postnatal head ultrasound underwent MRI brain except one with partial dysgenesis of corpus collasum on ultrasound. The abnormalities found on postnatal scans are shown in table-1. Postnatal head ultrasound and MRI brain are shown in fig-1. All cases with abnormal postnatal imaging studies 8/20 (40%) were followed in pediatric neurology clinic for development and other neurological morbidities. Neurological examination was normal in 5/8 cases (62.5%) including two with isolated MCM on postnatal MRI.

In 3/8 cases (37.5%) with associated CNS anomalies, there was mild motor delay with no major neurological sequalae. Speech and cognition was delayed in one case with cerebellar hypoplasia with normal motor development. Cases with partial dysgenesis of corpus callosum had mild developmental delay with normal eye contact and no signs of hypothalmic pituitary dysfunction

DISCUSSION

Isolated MCM is a rare CNS anomaly. Prenatal ultrasound and fetal MRI can identify this anomaly. It is crucial to exclude cerebellar hypoplasia prior to making this diagnosis. The reported incidence in literature is 1/8268 births[10]. In our study isolated MCM was confirmed in 2 cases (1/12,721 births) on postnatal neuro-imaging studies. We found male preponderance of MCM that is consistent with what has been reported in the literature[8,10].

Fetal sonography is the primary fetal imaging technique in our hospital. However, limitations with fetal sonography are already described in the literature with variable specificity and false positive diagnoses of posterior fossa anomalies[11-13]. False positive antenatal scans in our study were 60%. Limperopoulos et al found limitations in both sensitivity and specificity of fetal MRI and confirmed only 60% of prenatal fetal MRI findings on post natal MRI[14].

MCM as a part of complex CNS anomalies changes the prognosis and long term outcome. In our study postnatal neuro-imaging revealed

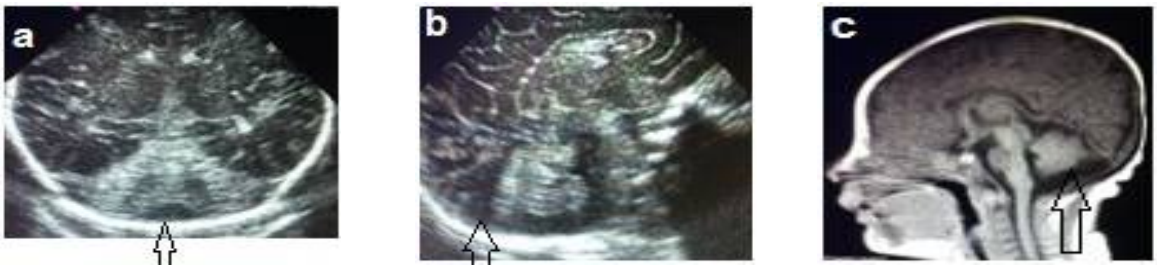

Figure-1: Coronal (a) and sagital (b) view of postnatal head ultrasound showing dilated cistern magna. Postnatal MRI brain (c) of same case showing prominence of cistern magna with some degree of cerebellar hypoplasia.

associated CNS anomalies in 6/20 (30%) cases with isolated MCM on antenatal fetal scan compared to 54.5% reported by Long et al[10]. In a series of 90 cases of suspected fetal posterior fossa anomalies by fetal sonography and 60 (67%) of them by fetal MRI, 42 were born alive and 39/42 had postnatal MRI, 10 cases (26%) had additional supratentorial malformations and more extensive cerebellar anomalies not evident on the fetal studies[14]. Colleoni et at found two cases with cortical malformations that had escaped prenatal diagnosis resulted in a worse outcome than had been predicted antenatally[15]. Clinical significance of isolated mega cisterna magna is not clear and most of the studies have reported a good prognosis.[3,6] Zimmer et al reported an overall normal cognitive function in adults but inferior score in some parameters of memory and verbal fluency compared to controls[16]. In our study the normal outcome was 50% in MCM cases with associated CNS anomalies compared to 81% reported by Wald et al[11]. Outcome is reported to be normal in 92% of survivors with isolated MCM without CNS malformations[11] while our 2 cases with isolated MCM have normal neurological outcome. However it is difficult to draw conclusion about neurological morbidities due to limitations of the standardized neurological evaluation in our cases.

We hope our study will help in counseling the parents about this rare anomaly especially in antenatal life where fetal ultrasound is used as screening tool. Limitations of our study include retrospective nature and all the antenatal fetal scan with MCM were not investigated in post natal period due to loss of follow up. Further prospective studies are warranted to know the incidence, associated CNS anomalies and neurological outcome. The fetal diagnosis of MCM should be investigated and followed up in our population due to associated CNS anomalies that may change the outcome.

CONCLUSION

Although, isolated MCM is not clinically significant but its morbidities are significant if associated with other brain malformations. MRI brain is a better modality for confirming and diagnosing associated malformations of the brain. MCM detected on fetal sonography needs to be interpreted with caution especially if done early in pregnancy. Follow up fetal and/or postnatal MRI is needed to rule out the associated CNS malformations.

CONFLICT OF INTEREST

The authors of this study reported no conflict of interest.

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