The Journal of International Medical Sciences Academy
2018 ; (January – March); Vol 31; No. 1

Case Report

Prenatal Diagnosis of Hemimegalencephaly: A case report with review of literature.

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Abstract

Prenatal diagnosis of hemimegalencephaly an extremely rare entity characterized by enlargement of all or portions of cerebral hemisphere.we report a case of unilateral megalencephaly with unilateral ventriculomegaly of the left cerebral hemisphere with unilateral ventriculomegaly. A woman presented at 30 weeks gestation for fetal ultrasound scanning and magnetic resonance imaging screening after an abnormal fetal ultrasound. Fetal magnetic resonance imaging showed hemimegalencephaly of the left cerebral hemisphere with unilateral ventriculomegaly.

Conclusions: In utero diagnosis of complex developmental brain anomalies allows a multidisciplinary approach to provide optimal prenatal patient treatment and parental counseling.

Introduction

Hemimegalencephaly (HME) is a congenital brain malformation characterized by the overgrowth and enlargement of all or parts of a cerebral hemisphere. It can rarely involve the brainstem or cerebellar hemisphere.

HME can occur as part of a genetic syndrome, such as epidermalnevus syndrome, Proteus syndrome, or tuberous sclerosis, or it can occur in isolation due to possible abnormality in cortical development and neuronal migration and possibly an early disturbance in cellular lineage and differentiation.

Radiologically, HME shows an enlarged unilateral cerebral hemisphere with abnormal lgyrationan and ventriculomegaly. USG and MRI are the imaging modalities.

Case report

Primi gravid was referred for antenatal scanning and showed-Fetal USG showed BPD 7.65cm - 30.5 wks, HC 28.9 - 31. 6wks, AC 255-29.5 wks, Fatty liver. 5.3-28,2 wks and average gestational age of 30.1 wks. Left lateral ventricle was 2.4 cm, dilated and right cerebral hemisphere width was less than the right cerebral hemisphere. Screening MRI showed ventricular asymmetry, midline shift with displacement of the occipital lobe across the midline, large dilatation mainly at the posterior horn of the left lateral ventricle, and a head circumference in the 90th percentile without involvement of the brain stem and cerebellum.

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Received : 13.02.19
Accepted : 12.03.19

Discussion

HME is a rare developmental brain disorder characterized by the abnormal enlargement of the entire cerebral hemisphere or portions of it and rarely brainstem [1,2]. HME can occur as an isolated case (genetic mutation) however or it can also occur with neurocutaneous and other similar syndrome like proteus, epidermal nevus, klipple trenaunay syndromes, TaySachs disease, mucopolysaccharidoses, Canavan disease.

The most common clinical presentation of affected postnatal patients is intractable epilepsy with severe developmental delay and psychomotor retardation typically starting from birth or within the first few months of life. HME is usually associated with agyrea, poly microgyrea [3].

Prenatal diagnosis with USG to be followed with MRI to assess tha associated anomalies. Prenatal CNS ultrasound and MRI as in the our case revealed ventricular asymmetry, midline shift with displacement of the occipital lobe across the midline, large dilatation mainly at the posterior horn of the left lateral ventricle, and a head circumference in the 90th percentile without involvement of the brain stem and cerebellum [4]. (fig. 1, 2)

Usg (fig.1) and MRI (fig.2) shows colpocephaly with hemimegalencephaly.

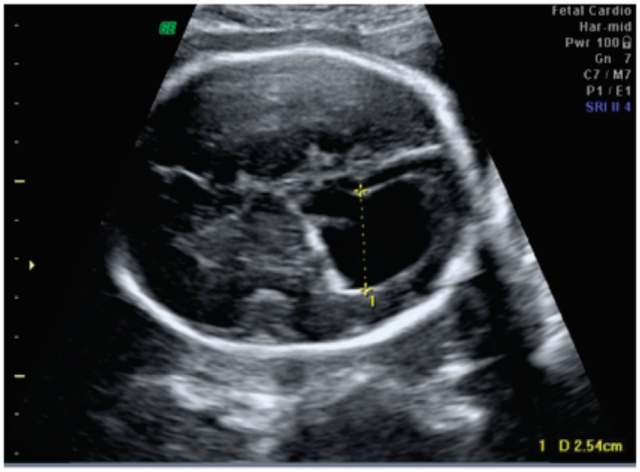

Fig.1: USG shows colpocephaly with hemimegalencephaly.

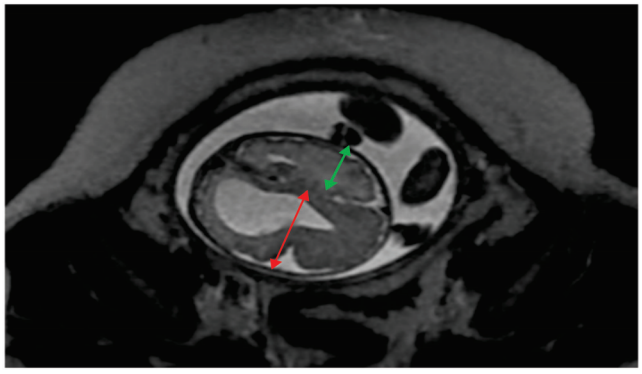

Fig. 2: MRI shows colpocephaly with hemimegalencephaly.

Conclusion

Unilateral megalencephaly is a rare anomaly of neuronal cell migration. We recently diagnosed this condition in a fetus at 20 weeks' gestation with ultrasonography and magnetic resonance imaging.

Acknowledgements

The authors wants to thank Dr.venugopal Medical SUPT., Dr. Krishna Rao,Principal, Dr. Roopa Suresh AMD, Dr. Suresh Krishna Murthy MD and Dr. Ramesh Kumar Prof & HOD

Conflict of interest:	All authors declare no COI
Ethics:	There is no ethical violation as it is based on voluntary anonymous interviews
Funding:	No external funding
Guarantor:	Dr. B. Mallikarjunappa will act as guarantor of this article on behalf of all co-authors.

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