

Case Report

OPEN LIP SCHIZENCEPHALY WITH AGENESIS OF CORPUS CALLOSUM: A RARE EMBRYOLOGICAL DEFECT IN COMBINATION

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ABSTRACT

A five week old newborn presented with recurrent abnormal movements. The child was borne of normal parents with non consanguineous marriage. There was no history of any ailment of mother during pregnancy and she was not on any drugs. The baby was borne by normal delivery and The CT Scan of the head showed Schizencephaly with Agenesis of Corpus Callosum with gyral malformation. There were no associated anomalies. Facial features were well developed and the muscle tone and power of the child was normal. The blood investigations were normal as was metabolic profile.

KEY WORDS: Callosum, Agenesis, Lateral Ventricle, Commissure, Forceps.

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Access this Article online

Quick Response code



DOI: 10.16965/ijar.2014.560

Web site: International Journal of Anatomy and Research
ISSN 2321-4287
www.ijmhr.org/ijar.htm

Received: 24 Dec 2014

Peer Review: 11 Dec 2014 Published (O):31 Jan 2015

Accepted: 20 Jan 2015 Published (P):31 Mar 2015

INTRODUCTION

The Corpus Callosum is an important commissure of Brain. It connects the two central hemispheres and maintains coordination between the two hemispheres. Its anomalous development is usually associated with other developmental defects. Schizencephaly is other developmental defect in which there is defective grey matter development. Both these conditions are not often found in combination. The article presents a rare case where both these conditions co exist.

DISCUSSION

Normal Anatomy: Corpus callosum is the largest & the chief commissure and contains

fibres which connect all areas of the two central hemispheres except the right and left temporal poles. It consists of rostrum, genu, body and splenium. The fibres which connect the prefrontal regions pass through the genu and anterior part of the body of the corpus callosum and make an arch as soon as they cross the middle line; this arch is called the Forceps anterior (minor).The fibres which connect the occipital poles pass through the splenium and the posterior part of the body. And to make an arch as soon as they cross the middle line; this arch is called Forceps posterior (major.)The fibres in the middle part (body) of Corpus callosum pass transversely but after they cross the middle line they pass upwards and down-

wards in the white matter of the hemisphere to reach the different areas of the cortex. The occipital part of the Corpus callosum forms the roof and lateral wall of the posterior horn of the lateral ventricle and is called the Tapetum. Agenesis of Corpus Callosum is seen in certain new borns and they cannot coordinate the activity of both sides [1,2]. This may be associated with Dandy Walker Syndrome, Chiari II Malformation Holoprosencephaly, Liopma of Corpus Callosum .The Embryological Development of Corpus Callosum occurs between 16-20 weeks. It needs proper migration. Schizencephaly is seen to appear as a CSF filled cleft extending into the lateral ventricle with surrounding Dymorphic grey matter [3]. The lateral ventricle is the cavity of the cerebral hemisphere. Each cerebral hemisphere contains a large "lateral ventricle" which tubular space shaped like the letter C. Each lateral ventricle has a body, an anterior, a posterior and an inferior horn. The anterior horn lies in the frontal lobe, the posterior horn in the occipital lobe and the inferior horn in the temporal lobe.

Fig. 1: Demonstrating Corpus Callosum Agenesis.

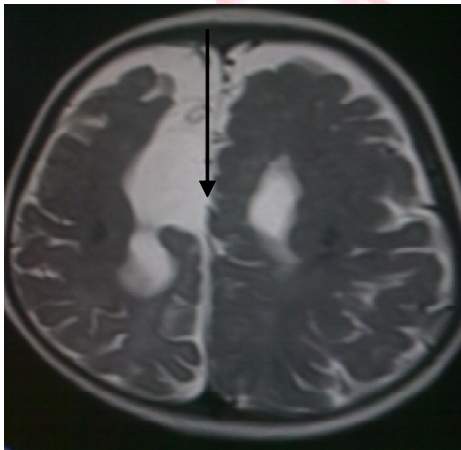


Fig. 2: Scizencephaly with absence of Well Defined Corpus Callosum.

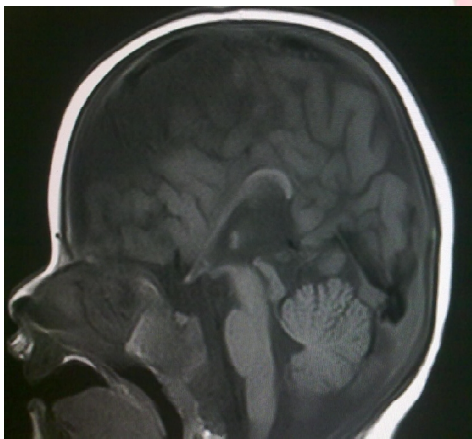
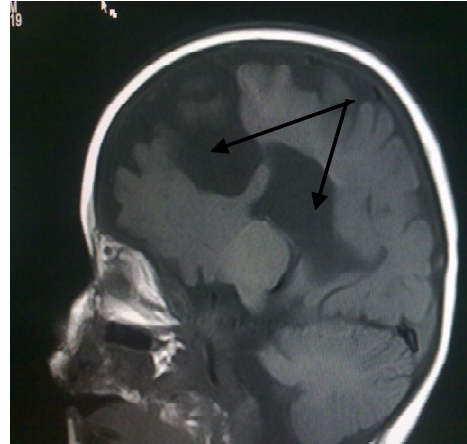


Fig. 3: Scizencephaly with absence of Corpus Callosum (Grey Matter Lined CSF Cleft Extending from Lateral Ventricle).



These clefts can be of varied sizes. The open lip type of schizencephaly as presented in this case has separate lips and cleft extending into lateral ventricles. It has been observed that a vascular defect during embryogenesis is the main cause. An infarction in the area of germinal matrix layer of the cerebrum during seventh week causes vascular compromise and the result is Grey Matter Lined CSF Cleft Extending from Lateral Ventricle. There may be added insult resulting from defective stem cell differentiation, concurrent metabolic defects and other cortical developmental defects like lissencephaly, paccygyria as well as polymicrogyria [4,5].

There may be multiple associated causations like infective factors, genetic factors, associated syndromes, mutations in genes. MRI appears to be the modality of choice because of ability to differentiate grey matter and white matter better. Associated agenesis of corpus callosum [6] is not an uncommon occurrence .It can result in incoordination , macrocephaly, visual impairment , mental retardation and seizures.it may e associated with Apert Syndrome, .Lowe Syndrome, Gorlin syndrome, Fryns Syndrome, Trisomies(Trisomy 18, Trisomy 13), Aicardi Syndrome, Shapiros Syndrome .

CONCLUSION

Though the combination of Open Lip Schizencephaly with Agenesis of Corpus Callosum is a rare event but the subsequent development of child with such defects are a source of multiple problems to the child in the form of Non Development, Mental Retardation,

Recurrent seizures with the added fact that mostly they are associated with other syndromes. As such they are a source of potential concern for the Child as well as Parents who find huge difficulties in raising such children should they survive.

Conflicts of Interests: None

REFERENCES

- [1]. Chayim Can Schell-Apacik, Kristina Wagner, Agenesis and Dysgenesis of the Corpus Callosum: Clinical, Genetic and Neuroimaging Findings in a Series of 41 Patients. *Am J Med Genet A*. 2008 October 1; 146A(19):2501–2511.
- [2]. Lynn K. Paul. Developmental malformation of the corpus callosum: a review of typical callosal development and examples of developmental disorders with callosal involvement. *J Neurodev Disord*. 2011 Mar;3(1):3-27.
- [3]. Oh KY, Kennedy AM, Frias AE, Byrne JLB: Fetal schizencephaly: pre- and postnatal imaging with a review of the clinical manifestations. *Radiographics* 2005;25:647-57.
- [4]. Barkovich AJ, Chuang SH, Norman D: MR of neuronal migration anomalies. *AJR Am J Roentgenol* 1988 Jan;150(1):179-87.
- [5]. Kuzniecky RI, Barkovich AJ. Pathogenesis and pathology of focal malformations of cortical development and epilepsy. [Review]. *J Clin Neurophysiol* 1996; 13: 468–80.
- [6]. Del Campo M, Hall BD, Aeby A et al. Albinism and agenesis of the corpus callosum with profound developmental delay: Vicisynndrome, evidence for autosomal recessive inheritance. *Am J Med Genet* 1999;85:479-485.

How to cite this article:

Ashfaq ul Hassan, Shifan Khanday, Aijaz Rather, Muneeb ul Hassan. OPEN LIP SCHIZENCEPHALY WITH AGENESIS OF CORPUS CALLOSUM: A RARE EMBRYOLOGICAL DEFECT IN COMBINATION. *Int J Anat Res* 2015;3(1):838-840. **DOI:** 10.16965/ijar.2014.560

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