



Cephalic Disorders with Neuro-Ophthalmic Manifestations

Jinat SF^{1*}, Chhara TR¹, Sibgatullah M², Nahar N³ and Jafrullah TA⁴

¹Assistant Professor and Consultant, Ispahani Islamia Eye Institute and Hospital, Bangladesh

²Associate Professor and Senior Consultant, Ispahani Islamia Eye Institute and Hospital, Bangladesh

³Professor and Senior Consultant, Ispahani Islamia Eye Institute and Hospital, Bangladesh

⁴Consultant, Green Eye Hospital, Bangladesh

Case Report

Volume 7 Issue 1

Received Date: April 25, 2022

Published Date: May 10, 2022

DOI: 10.23880/oajo-16000241

*Corresponding author: Syeda Fatima Jinat, Assistant Professor and Consultant, Ispahani Islamia Eye Institute and Hospital, Sher e bangla ngor, Farmgate, Dhaka-1215, Bangladesh, Tel: 01711480570; Email: syeda.fatima@islamia.org.bd

Abstract

Purpose: To report three different cases of cephalic disorders with neuro-ophthalmic manifestations.

Method: Descriptive case report.

Case Summary

Case 1: 14 years female came with Primary optic atrophy (POA) of both eyes associated with poor intelligence. MRI brain showed disproportionate dilatation of both occipital horn of lateral ventricle suggestive of Colpocephaly.

Case 2: 16 years male presented with POA of eyes, horizontal jerky nystagmus and left sided hemiparesis with contracture of left hand since birth. MRI brain suggestive of Open lip Schizencephaly.

Case 3: 12 years boy came with alternate exodeviation, left sided hemiparesis since birth. MRI brain suggestive of right sided Procencephalic cyst.

Conclusion: Cephalic disorders caused by disturbance in development of fetal nervous system. For most cephalic disorder's treatment is only symptomatic and supportive. Early diagnosis may ensure patient's quality of living.

Keywords: Neuro-Ophthalmic; Cephalic Disorders; Primary Optic Atrophy; Open lip Schizencephaly; Procencephalic cyst

Introduction

Cephalic disorders are congenital condition that stem from damage to, or abnormal development of the budding nervous system. More common in premature baby specially those born with hypoxia or lung immaturity. It is caused by hereditary or genetic conditions or by environmental exposure during pregnancy such as medication, maternal infection or exposure to radiation [1].

In this study, we will report 3 different cases of cephalic

disorders along with neuro-ophthalmic manifestations.

Case No 1

A 14 year female came with poor vision of both eyes since childhood. She was dumb with poor intelligence. Vision in right eye was 6/36, left eye 6/24, not improved with pin hole. Color vision was impaired in both eyes, pupillary reaction in both eyes were sluggish, both the optic discs were pale (Figure 1).

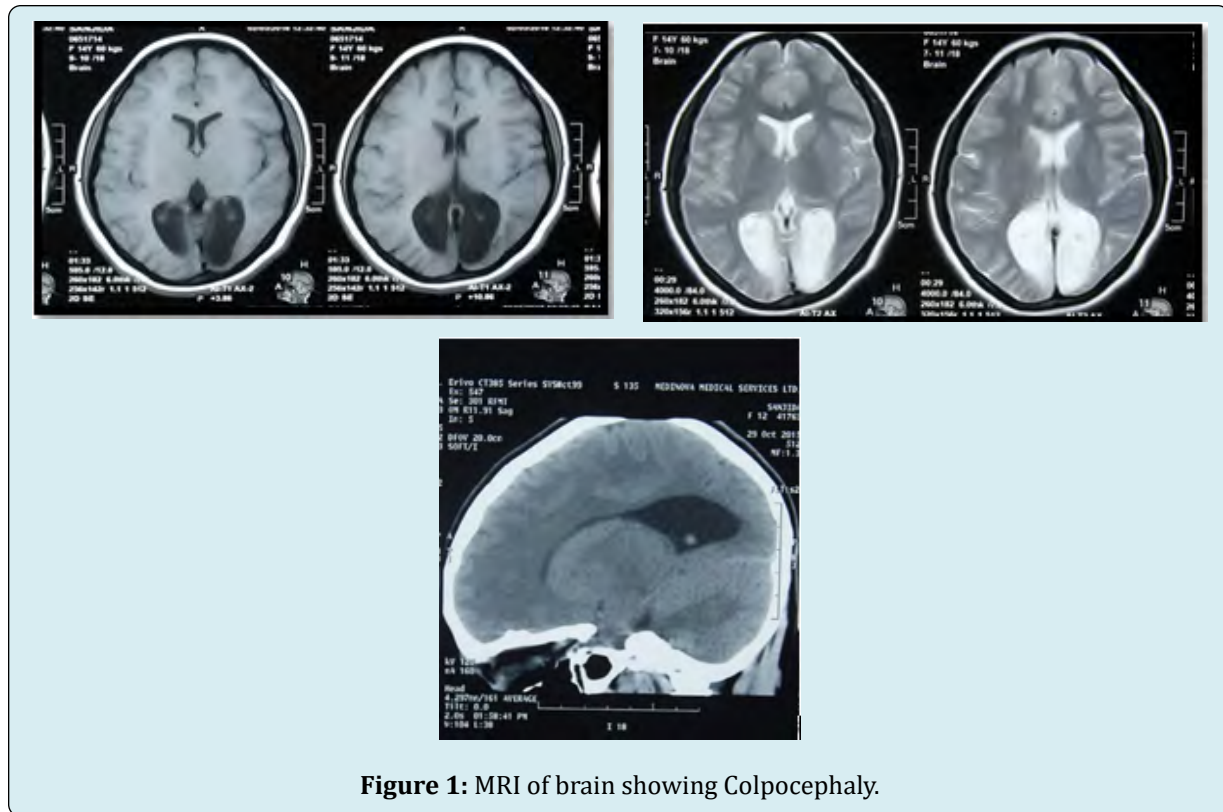


Figure 1: MRI of brain showing Colpocephaly.

MRI of brain showed disproportionate dilatation of both occipital horn of lateral ventricle with hypoplastic corpus callosum, feature suggestive of Colpocephaly.

Case No 2

A 16 year male came with poor vision of both eyes along

with weakness of left side of the body since childhood. Visual acuity in both eyes were 6/36, not improved with pinhole, color vision was significantly impaired, pupillary reaction in both eyes were sluggish, optic discs were pale and there was horizontal jerky nystagmus. The patient had left sided hemiparesis with deformity of left hand (Figure 2).

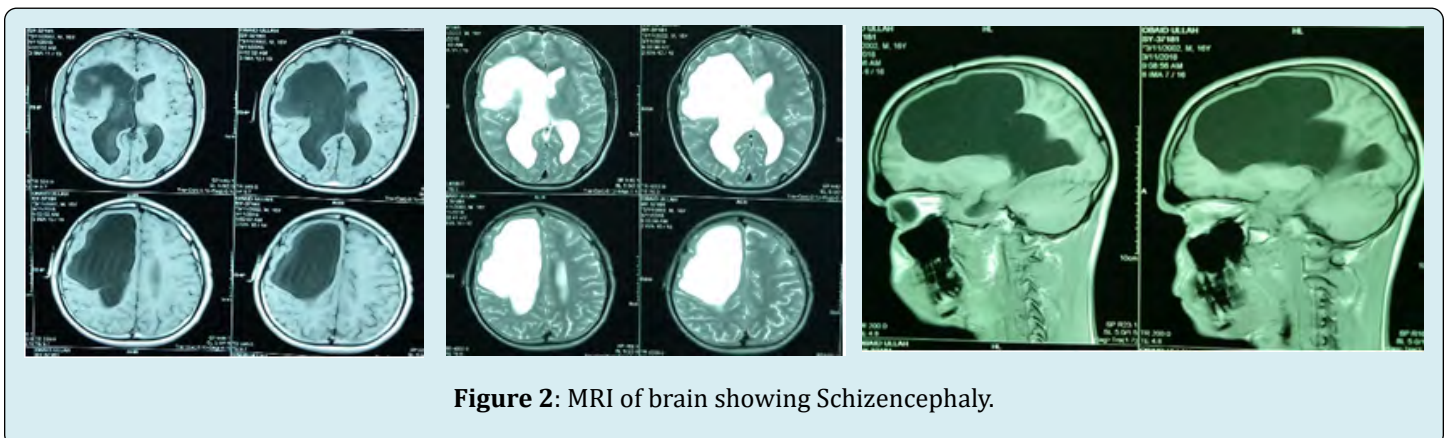


Figure 2: MRI of brain showing Schizencephaly.

MRI of brain showed a large CSF intensity lesion involving right fronto-parietal lobe communicating to right lateral ventricle with moderate dilatation of both lateral ventricles, feature suggestive of Open lip Schizencephaly.

Case No 3

A 12 year boy presented with alternate outward deviation of both eyes, weakness of left side of the body since birth. There was alternate exodeviation, visual acuity in right

eye was 6/6 and left eye 6/9, not improved with pin hole and left sided hemiparesis. Both the fundi were normal (Figure

3).

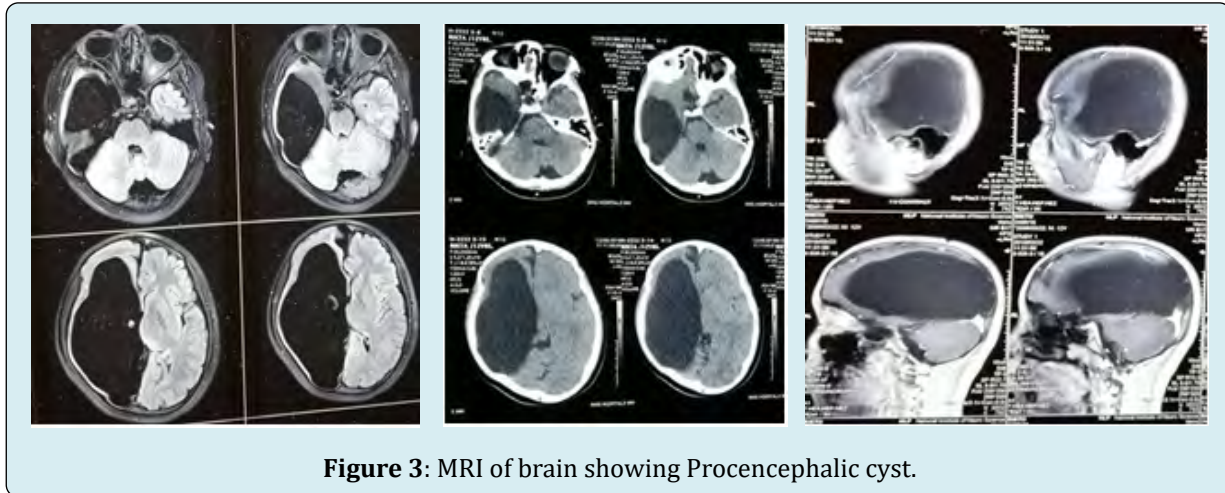


Figure 3: MRI of brain showing Procencephalic cyst.

MRI of brain showed CSF filled large cavity in right cerebrum communicating with right lateral ventricle with thinning of overlying cerebral parenchyma, feature suggestive of Procencephalic cyst.

Discussion

Colpocephaly believed to occur as a result of neuronal migration disorders during early brain development, intrauterine disturbances, perinatal injuries and other central nervous system disorders and may present with various degrees of motor disabilities, visual defects, spasticity and moderate to severe intellectual disability [2].

Schizencephaly is a rare congenital malformation with prevalence of 1.48 in 100,000 live births [3]. The defect characterized by abnormal clefts or slits in cerebral hemisphere of brain which may be unilateral or bilateral, lined with grey matter. Common clinical features include microcephaly, hydrocephalus and intellectual disability, partial or complete paralysis and or hypotonia [4].

Procencephalic cyst is an extremely rare cephalic disorder, characterized by congenital or acquired cavity within cerebral hemisphere, containing cerebrospinal fluid [5]. It occurs due to disturbance of vascular supply leading to cerebral degeneration, also due to abnormal development (malformative), direct damage, inflammation or haemorrhage [6]. The patient may suffer from minor neurological deficit to severe disability even death.

Conclusion

The cause of cephalic disorder is multifactorial and neuro imaging is the investigation of choice. The condition is

usually nonfatal. There is no definitive treatment and visual prognosis is guarded. If physiotherapy, occupational therapy, speech therapy given in early state, may reduce morbidity and improve quality of living.

Conflict of Interest

There is no conflict of interest involved.

References

1. Cephalic Disorders Fact Sheet. The National Institute of Neurological Disorders and Stroke (NINDS).
2. Puvabanditsin S, Garrow E, Ostrerov Y, Trucanu D, Ilic M, et al. (2006) Colpocephaly: a case report. *Am J Perinatol* 23(5): 295-297.
3. Barkovich AJ, Norman D (1988) MR imaging of schizencephaly. *AJR Am J Roentgenol* 150(6): 1391-1396.
4. (2014) NINDS Schizencephaly Information Page. National Institute of Neurological Disorders and Stroke.
5. Parker J (2004) The official parent's sourcebook on porencephaly: A revised and updated directory for the internet age. ICON Health Publications.
6. Debus O, Kosch A, Strater R, Rossi R, Nowak-Gottl U (2004) The Factor V G1691A Mutation is a Risk for Porencephaly: A Case-control Study. *Annals of Neurology* 56(2): 287-290.

