

BILATERAL PRIMARY OPTIC ATROPHY IN A PATIENT WITH HYDROCEPHALUS – A RARE CASE

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ABSTRACT

Objective – To report the rare ophthalmological findings in a patient diagnosed with hydrocephalus

Material And Methods – We report the case of a 2yr old female child who presented with hydrocephalus. CT Brain was done which was suggestive of gross dilatation of both the lateral ventricles and fourth ventricle and thinning of the cortex was seen. A detailed ophthalmological evaluation was done which showed reduced ocular motility, lagophthalmos and right esotropia. Fundus examination showed bilateral primary optic atrophy with blurred margins of the optic disc. **Results** – The clinical history, detailed ophthalmological examination and investigations suggest that hydrocephalus and thinning of the cortex led to retrograde optic atrophy in this patient. **Conclusion** – Hydrocephalus and thinning of the cortex led to retrograde optic atrophy. Due to high degree of thinning of the occipital lobe caused by hydrocephalus, we do not expect the patient's visual functions to develop in future. Awareness regarding consequences of untreated cases of hydrocephalus and screening for ophthalmological signs is important. Timely intervention can prevent progression of the disease and lead to improved quality of life in such patients.

KEYWORDS : Hydrocephalus Thinning of cortex Primary optic atrophy

Case Report –

We report a 2yr old female child born out of a consanguineous marriage at gestational age of 7 months. The mother gives history of having developed preeclampsia and seizures in the 7th month of gestation after which the baby was delivered by normal vaginal delivery. The birth weight of the baby was 1kg. She was admitted to the NICU and was treated with surfactant and oxygen therapy for 1.5 months after which she was discharged. Her weight at the time of discharge was 2.5kg.

After 5 months the parents noticed unusual enlargement in the head circumference of the baby when she was diagnosed with hydrocephalus and patient underwent a surgery for ventriculo-parietal shunt. The patient showed delayed milestones. Has no history of seizures. The patient was vaccinated till date.

After 1.5 years the parents again noticed unusual enlargement of the head circumference of the baby and consulted at a private hospital. CT brain was performed which was suggestive of gross dilatation of both the third ventricles and the fourth ventricle as well as thinning of the cortex was seen. The patient underwent a surgery for parietal external ventricular drain and removal of the non-functioning VP shunt with excision of brain abscess. Following this the patient was admitted at our hospital for post operative care.

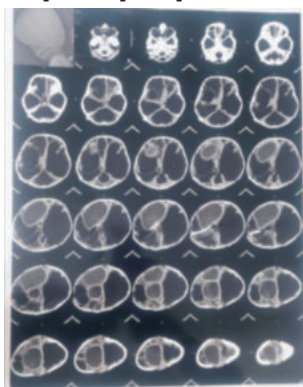


Fig 1. CT Brain showing gross dilatation of both the third

ventricles and the fourth ventricle as well as thinning of the cortex

We performed a detailed ophthalmological examination and the ophthalmological features noted in this patient were as follows –

- There was reduced blinking
- Reduced ocular motility.
- Lagophthalmos was noted.
- The patient did not fixate or follow light or objects.
- Right esotropia was seen.
- Hirschberg test showed a deviation of 15° in the right eye
- No abnormal anterior segment findings were observed such as pupil abnormalities, cataract, nystagmus or ptosis.
- **Fundus examination** of both eyes indicated **primary optic nerve atrophy** with blurred disc margins, but the degree of pallor was mild. The colour of the fundus was weak and of reddish tone yet no abnormality was found in the diameter and development of the retinal vessels which normally extend to the peripheral portion of the fundus. No obvious chorioretinal dysplasia was seen.



Fig. 2 Clinical picture of the patient

DISCUSSION –

Hydrocephalus (HC) is an enlargement of the head which is caused by an accumulation of the cerebrospinal fluid (CSF). In order to relieve the pressure caused by this accumulation,

surgery is necessary to elevate ventricular volume and pressure to safe levels. The medical device used to remove extra CSF and restore the normal pressure and flow is called a ventriculoperitoneal (VP) shunt.^[1] Many ophthalmological complications, including difficulties with ocular motility, defects in the visual field, optic atrophy, and loss of visual acuity, have been attributed to hydrocephalus.^[2]

Further ocular motility symptoms, such as convergent squints, lateral rectus palsy, and divergence palsies, can point to VP shunt malfunction and elevated intracranial pressure.^[3,4] Also, it has been claimed that ocular motility disorders and visual function are significantly impacted by the age at which hydrocephalus first manifests, rather than the cause of the condition, the quantity of shunt revisions, or the size of the ventricle.^[5]

The pathological term "optic atrophy" characterizes the shrinking of the optic nerve brought on by the axonal degeneration of retinal ganglion cells (RGCs). The injury to the visual pathway is characterised by optic atrophy.^[6] Optic nerve atrophy is not a disease; rather, it is a warning sign that may indicate a more severe condition to the ophthalmologist. Therefore, a comprehensive examination of a child who is presenting with optic atrophy is essential, and being aware of the common causes helps the physician conduct the proper work-up and administer the necessary treatment. This is crucial for children because early detection and treatment of a progressive or life-threatening condition may prevent systemic morbidity and vision impairment in later years, or else direct the family towards the proper visual, scholastic, and social recovery.^[7]

CONCLUSION –

Hydrocephalus and thinning of the cortex led to retrograde optic atrophy in this patient. Due to high degree of thinning of the occipital lobe caused by hydrocephalus, we do not expect the patient's visual functions to develop in future. Awareness regarding consequences of untreated cases of hydrocephalus and screening for ophthalmological signs is important. Timely intervention can prevent progression of the disease and lead to improved quality of life in such patients.

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Conflict Of Interest –

There is no conflict of interest.

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