Is it Hydranencephaly - A Variant?

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Introduction

Hydranencephaly is a rare devastating malformation where greater part of the cerebral hemisphere is replaced by large membranous sac filled with cerebrospinal fluid (CSF) [1,2]. Similar condition can be produced secondary to a congenital/perinatally acquired hydrocephalus. Both these conditions mimic each other on CT and are trans-illumination positive. Literature search does not give any definite clinical criteria to distinguish these two. We therefore present a similar case, which posed a diagnostic problem.

Case Report

A 3 month old male infant, product of nonconsanguinous marriage was born at 36 weeks of gestation with a birth weight of 1.5 kg. Infant was admitted at 3rd day of life with features suggestive of sepsis and treated with parenteral antibiotics for two weeks. At 6 weeks of age he was detected to have rapidly increasing head circumference. Examination revealed occipito frontal circumference (OFC) of 39 cms. Trans-illumination was strongly positive. He had hypotonia with hypeflexia. CT scan revealed membrane filled sacs compressing cerebral hemisphere to a thin rim.

Discussion

A variety of destructive developmental abnormalities occurring after the fourth month of gestation may lead to hydranencephaly [3]. There is some evidence that it may arise as a result of obstruction to blood supplied by the internal carotid artery (ICA) in fetal life occurring after the fourth month of gestation [2,3]. Although it is generally accepted that this condition occurs secondary to an in-utero occlusion of ICA, maternal cocaine, CMV and Toxoplasmosis are also implicated [3]. Postnatal causes of hydranencephaly are meningitis, intracerebral infarction and ischaemia, which also cause obstruction to CSF flow because of the effects of inflammatory process, leading to creation of a setting for hydrocephalus.

In these two conditions one is at loss to distinguish between severe obstructive hydrocephalus with extensive cerebral parenchymal damage and hydranencephaly. Extensive loss of cerebral parenchyma because of compression by CSF filled spaces, being a common denominator. In such a situation there exists a need for laying down criteria for distinguishing the two.

In our case the child was normal at birth and developed increasing OFC with positive trans-illumination test at 15 days of life. CT scan revealed fluid filled sacs in both hemispheres R>L with dilated 3rd ventricle and extensive destruction of cerebral parenchyma in the anterior cranial fossa (frontal lobe) reducing it virtually to a rim of cortex. This satisfies the diagnosis of obstructive hydrocephalus and also conforms to the universally accepted definition of hydranencephaly.

As on today, MRA is the only definitive diagnostic modality to differentiate between hydranencephaly and-

Fig. 1: Increased OFC at 6 weeks

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associated with enlarging head and hydrocephalus [4]. In the CT picture of our case, there is an absence of frontal lobe parenchyma leading to grossly dilated frontal horns of lateral ventricle, which is virtually occupying the entire anterior cranial fossa. Absence of parietal lobe with its parenchyma being replaced by grossly distended body of lateral ventricle is also visible. On the other hand temporal lobe parenchyma is visible with normal size temporal horn of the lateral ventricle.

In view of the above, it appears that the occlusive process has involved the anterior CA completely and middle CA only partially. Obstructive hydrocephalus with site of obstruction at aqueduct of sylvius would have resulted in dilatation of third ventricle with involvement of both horns of lateral ventricle, which is not seen in this case. It is therefore logically reasonable to deduce that it is at best a variant of hydranencephaly with complete involvement of anterior CA, while involvement of medial CA has been partial.

Differentiating between these two conditions ie hydranencephaly and obstructive hydrocephalus is of great importance as regards prognostication, therapeutic intervention and counselling of the parents as hydranencephaly can be autosomal recessive [3] and needs genetic counselling. Shunt does not have any beneficial role in hydranencephaly. On the other hand hydrocephalus has no definite genetic mode of transmission and shunt does have a beneficial role. Therefore authors feel that definite criteria be laid down for distinguishing these two conditions.

References

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Define a baby - "A loud noise at one end and no sense of responsibility at the other."

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In General

Behind every successful man there is a woman. However don't you think that in these competitive times there is need for more than one woman behind every successful man?!!!!!