Case Report

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Benign External Hydrocephalus

Thaslima K.*, Sunil Mhaske, Vishnu Kadam*****

*Resident, ** Professor & Head, ***Associate Professor, Department of Paediatrics, PDVVPF's Medical College, Ahmednagar, Maharashtra.

Abstract

The term external hydrocephalus or benign enlargement of subarachnoid space (BESS)was first used by Dandy in 1917 to describe enlargement of the subarachnoid space in the presence of increased intracranial pressure [1]. External hydrocephalus is defined as a rapid increase in head circumference in an infant combined with enlarged frontal subarachnoid spaces as seen on CT, MRI, or cranial ultrasound and with normal or slightly enlarged ventricles [2].

It is more common in males and is mostly idiopathic but can be due to delayed development or delayed function of arachnoid villi at sagittal sinus, intraventricular and subarachnoid haemorrhage, prematurity, meningitis and trauma. Patient presents with a rapidly enlarging head, or with delay in gross motor development, hypotonia, or it can be an accidental finding on investigations. It is a normal variant of hydrocephalus and the most common cuse of macrocephaly [3].

External hydrocephalus is diagnosed by an ultrasound with MRI and CT scan being more diagnostic. There is an increased risk of subdural haematoma communicating hydrocephalus or subdural haematoma.Studies show that infants with macrocephaly or rapid head-growth, CT findings of enlarged subarachnoid spaces, normal-to-minimally increased ventricular size and who have a parent with macrocephaly, have a good developmental prognosis and a characteristic pattern of neuromotor development in the first year [4]. In this poster we discuss a case of benign external hydrocephalus in a 11month male child presenting with a history of head injury.

Keywords: Hydrocephalus; Macrocephaly; Subarachnoid Space.

Introduction

Hydrocephalus is a relatively common neuropediatric condition, with an incidence of about 0.9 per 1,000 births.It is defined as the abnormal accumulation of cerebrospinal fluid (CSF) within the ventricles and/or subarachnoid spaces, leading to an increase in intracranial pressure (ICP) [5]. Raimondi defined it as an increase in CSF volume.

It occurs mainly during infancy, and the subarachnoid space enlargement gradually decreases and disappears over the next year. The word "benign" is often used together with "external hydrocephalus," reflecting the common view that this is a self-limiting condition occurring during infancy, resolving spontaneously during childhood. Hence, most patients are probably not treated.

Case History

11 months male child visited the OPD with H/O gradually increasing head circumference since the

Corresponding Author: Thaslima K., Resident, Dept. of Paediatrics, PDVVPF's Medical College, Vilad Ghat, Ahmednagar, Maharashtra 414111.
E-mail: thaslima.k@gmail.com

past 1 month.His head circumference was more than 95th centile fort hat age.The anterior fontanelle was soft and of normal tension. Frontal bossing was present. Rest of the clinical examination revealed no significant abnormality. The child was neurodevelopmentally normal with normal milestones. The child was born at 39 weeks of gestation by normal vaginal delivery and had cried immediately after birth. Ct scan revealed subdural hygroma. Later an MRI was done showing benign enlargement of subarachnoid space. As thee was no neurological involovement patient was adviced follow up with MRI at 18-21months of age. He was given only conservative treatment as BESS is a self-limiting illness.

Fig. 1: Child with BESS syndrome

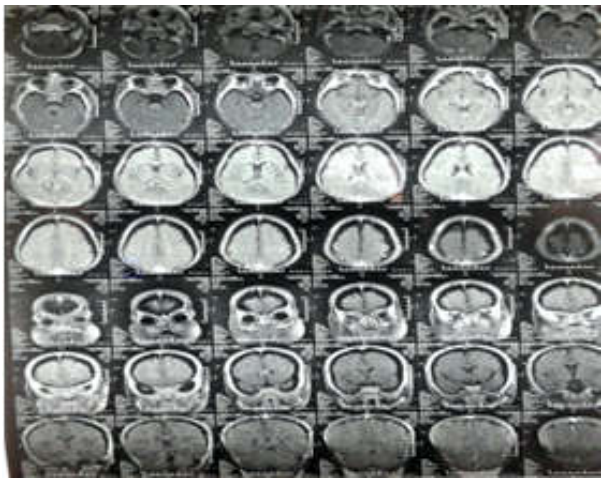

Fig. 2: MRI showing benign enlargement of subarachnoid space

Etiology

In most reported cases, there is no obvious cause of the external hydrocephalus, and it is therefore classified as idiopathic. However, it has been reported after numerous situations and conditions such as prematurity and intraventricular hemorrhage , meningitis, metabolic disorders , steroid therapy chemotherapy, neurosurgery , and trauma [6].

A complicating fact is that intraventricular and subarachnoid hemorrhages in premature infants often occur without symptoms , thus making it difficult to know if idiopathic hydrocephalus really is idiopathic or simply caused by such silent, clinical events .

External and communicating hydrocephalus is described in children with raised venous pressure, e.g., following various thoracic/cardiac conditions [7].

Heredity

Some patients with external hydrocephalus seem to have a familial form as one or more close relatives are macrocephalic.

An autosomal dominant mode of transmission has been assumed, although a multifactorial model of inheritance is the most recent proposal. The dominant inheritance might be due to a single gene exhibiting a major effect as part of a multifactorial phenomenon in some families , probably during a limited time of susceptibility in fetal development. Maytal et al. suggested that the primary phenotype merely was the delayed maturation of the arachnoid villi.

Pathophysiology

External hydrocephalus is caused by immature arachnoid villi not able to absorb the CSF that is produced continuously . The accumulated CSF then expands the ventricles and the subarachnoid space inside the compliant and growing skull of an infant, thus avoiding a marked increase in intracranial pressure. The arachnoid villi mature at about 18 months of age, ending the CSF accumulation and thus the widening of the subarachnoid space. Why the arachnoid villi do not mature remains unknown, but some heredity has been described [8]. The underlying mechanism for the formation of external hydrocephalus is poorly understood, although several theories exist. The familial macrocephaly associated with some of the cases indicates that heredity may play a role. CSF flow studies have shown reduced flow over the cerebral convexities; an impairment of CSF absorption through the arachnoid villi therefore seems intuitive. In normal children, it has been shown that the arachnoid villi are not fully mature at birth but that they gradually become so during infancy. This lack of maturation in combination with the pronounced increase in CSF production during the first year of life may be the

underlying mechanism and may also explain why the head starts to grow at around 6 months of age in most cases. This may not be a problem in most children, as their draining capacity through the villi or other draining pathways is balanced against the CSF production [8]. In children with external hydrocephalus, on the other hand, there may be a misbalance because of either delayed maturation or excessive CSF production.

Cerebrospinal Fluid Outflow

External hydrocephalus is commonly classified as a communicating hydrocephalus .Three pathways are recognized: the arachnoid granulations, the lymphatic capillaries, and the transependymal passage.

The term external hydrocephalus was first used by Dandy in 1917 to describe enlargement of the subarachnoid space in the presence of increased intracranial pressure. It was only recently that Robertson and Gomez reintroduced the term to describe a conditionin which children wtih enlarging heads have a CT scan of enlarged subarachnoid spaces with mild to moderate or no ventricular dilation. There appears to be an excess of normal CSF in the subarachnoid space. This was demonstrated by Andersson et al. who performed craniotomies on four patients with idiopathic EH and found an enlarged subarachnoid space without other abnormalities [9].

Risk Factors

External hydrocephalus may coexist with a series of conditions, such as some types of craniosynostoses, achondroplasia, Sotos syndrome, and glutaric aciduria type 1. A case of external hydrocephalus in a microcephalic infant has also been reported. The hydrocephalus in craniosynostosis and achondroplasia is supposedly caused by a rigid venous outflow obstruction.

Increased head circumference is found in all patients with external hydrocephalus. In most cases, the head circumference increases disproportionally only during the first year of life, an observation that may support the delayed maturation theory as discussed above. However, as the cranial sutures close between 1 and 2 years of age, it is difficult to exclude a persistently increased ICP. Many children end up with large heads, i.e., they to not normalize, signifying a continued growth stimulus beyond infant age [10].

A relatively common sign is a tense anterior fontanel. Other early symptoms and signs have also been reported occasionally: dilated scalp veins, frontal bossing (an unusually prominent forehead), irritability, hypotonia, vomiting, gross motor delay, ataxia, poor head control, seizures, fever, and mental retardation. We have not found any articles reporting sunset gaze.

Head Circumference

Infants with external hydrocephalus usually show a rapid increase in head circumference , which appears to be the most common symptom in all children developing hydrocephalus during their first year of life. Most of the increase in head circumference occurs around the age of 6. It seems that the head circumference usually stabilizes before the age of 18 months . Measurements afterwards typically lie above but parallel to the upper (95th to 98th) percentile. The amount of children ending up with macrocephaly varies considerably from 11% to 87% on long-term follow-up [11].

Hanlo et al. showed in a study of hydrocephalic infants that raised ICP is related to developmental outcome through the process of myelination as seen on MRI . Moreover, most children with severely delayed preoperative myelination showed at least a partial recovery following CSF diversion. The importance of myelination is supported by an animal study finding that white matter blood flow seems vulnerable in hydrocephalic kittens.

Investigations

Ultrasound, CT and MRI imaging of the brain may all demonstrate the characteristic findings seen in BESS:

- widening of the bifrontal and anterior interhemispheric CSF spaces
- no consensus of cut-off values exists
- findings should be correlated with patient age
- an estimated equation for both inter hemispheric width (IHW), craniocortical width (CCW) and sinocortical width (SCW) has been suggested
- IHW >5 mm in neonates
- IHW >8.5 mm in 1 year olds
- no flattening of adjacent gyri
- CSF space follows the gyral contour
- usually normal sulci posteriorly
- the anterior fontanelle is frequently enlarged with the enlargement of the subarachnoid space in the

frontoparietal regions

- normal ventricular size, no pressure effects on the surrounding brain tissue, no cerebral atrophy.
- no blood products on MRI study
- another key distinction between benign enlargement of the subarachnoid spaces and a subdural fluid collection is that in the former the cortical veins will be adjacent to the inner table of the calvarium on MR and ultrasound; whereas in the latter the veins are displaced away from the inner table, as the arachnoid membrane and subarachnoid space are displaced [12].

Complications

While these findings are benign in many cases, there is an increased risk of subdural haemorrhage, either spontaneously or following minor trauma. Subdural hematoma in a patient with BESS should not be interpreted as suggestive of non-accidental injury without other stigmata [13].

A low percentage of patients may develop communicating hydrocephalus, which may warrant treatment.

The condition resolves spontaneously by the age of 2 years. Although the macrocephaly may persist, the subarachnoid space fluid collection will resolve or become minimal as the child grows older.

Treatment

Studies show that infants with macrocephaly or rapid head-growth, CT findings of enlarged subarachnoid spaces, normal-to-minimally increased ventricular size and who have a parent with macrocephaly, have a good developmental prognosis and a characteristic pattern of neuromotor development in the first year [14].

A review of literature has described that although most children with external hydrocephalus do well, a substantial number show temporary or permanent psychomotor delay.

Conclusion

Although it is a self limiting disease and it usually resolves by 2 years of age, in some cases shunt operation is indicated. A review of literature has described that although most children with external hydrocephalus do well, a substantial number show temporary or permanent psychomotor delay. Hence, future research should focus on this, comparing the outcome of surgical treatment and conservative management of external hydrocephalus.

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