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Benign Enlargement of Subarachnoid Spaces in Infancy with Developmental Delay- A Case Report

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ABSTRACT

Background: Benign enlargement of subarachnoid spaces (BESS) is a common cause of macro crania in infancy and may present with developmental delay due to enlargement of the brain. The self-limiting nature of this condition allows most children to regain developmental milestones as they grow older. The aetiology of developmental delay in full-term infants with BESS is unclear and prognosis varies since only some children have been reported to have long-term deficit in gross motor and language domains.

Case presentation: This case report illustrates the clinical profile and findings on brain imaging in a full-term infant who presented with BESS along with delay in attaining age-appropriate gross motor and language milestones. Assessment of developmental milestones at 15 months of age supported the benign nature of this condition although the head circumference remained above the 95th percentile for age.

Conclusions: Despite the self-resolving nature of this condition, regular evaluation of developmental milestones is necessary to ensure timely intervention in appropriate cases. Further research on the aetiology could also aid clinicians in determining which children are at risk for developing long-term developmental delay.

Keywords: Benign enlargement of subarachnoid spaces; External hydrocephalus; Developmental delay; Macrocephaly; Case report

BACKGROUND

Benign enlargement of subarachnoid spaces in infancy is a self-limiting condition that presents as macrocephaly. It is more common in male infants with an enlarged head circumference in the father.[1] Head circumference at birth is usually normal and macrocephaly is evident at about 3-12 months of age.[2] These children lack features of raised

intracranial pressure or periventricular lucency although mild ventricular enlargement may be present.[3] It is also referred to as benign external hydrocephalus or extra-ventricular hydrocephalus.[2] Developmental delay in a child with BESS has been attributed to the stagnant flow of CSF in the subarachnoid space which affects cortical development since CSF helps in transporting growth factors and removing toxic metabolites. [4] While some children show normal development, others develop deficits in gross motor and rarely, language skills. Due to the benign, self-resolving nature of this condition, most children regain age-appropriate developmental milestones at follow-up. Developmental delay in children with BESS needs to be distinguished from other possible causes like TORCH infections, mucopolysaccharidoses, rickets and intracranial haemorrhage.

CASE PRESENTATION

A 11-month-old boy was brought by his parents to the out-patient department (OPD) with complaints of rapid increase in the size of his head over the past five months. He was born at term via lower segment caesarean section due to prolonged labour and weighed 3.5 kg at birth. Head circumference at birth was normal at 35 cm. Maternal and paternal head circumference was 53 cm and 55.5 cm respectively. Family history revealed no similar occurrence of macro crania. Prenatal history was insignificant for toxoplasmosis, rubella, cytomegalovirus, herpes simplex, and HIV (TORCH) infections during pregnancy. Postnatally, the child had no history of significant infection or trauma. Immunisations were up to date. Length and weight on presentation were 70 cm (between the 3rd-95th percentile) and 7.08 kg (<3rd percentile). Head circumference was 51 cm (>95th percentile for age) and examination revealed an enlarged, non-bulging and non-tense anterior fontanelle. Developmental assessment revealed delay in attaining gross motor milestones (developmental quotient (DQ) = 55) and a delay in speech (DQ = 64). On neurological examination, muscle tone, power and reflexes were within normal limits. Magnetic resonance imaging (MRI) revealed enlargement of subarachnoid spaces bilaterally in the frontal and temporal regions of the brain (Figure 1). A coronal section at the level of the interventricular foramen of Monro revealed a cranio-cortical width of 8.3 mm, an inter-hemispheric distance of 10.1 mm and a sino-cortical width of 11.4 mm. The ventricles, cortical sulci and basal cisterns were normal on imaging. There were no signs of pressure effect on the surrounding brain or cerebral atrophy. Thus, we arrived at a diagnosis of BESS. Follow-up evaluation at 15 months of age revealed a head circumference of 51 cm (>95th percentile) and a DQ of 80 and 67 on assessment of gross motor milestones and speech, respectively. Gross motor milestones and speech were regained to age-appropriate levels on evaluation at 18 months of age, however, the child's head circumference remained slightly above the 95th percentile.

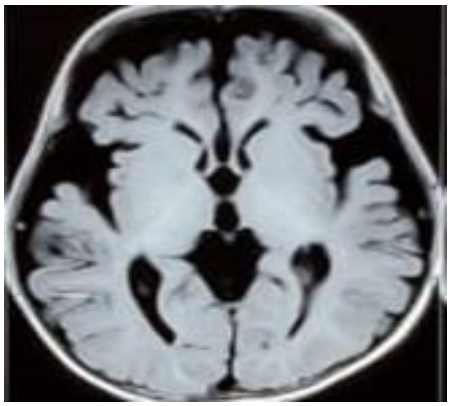

Figure 1: Axial T-1 weighted MRI showing enlargement of subarachnoid spaces in the frontal and temporal regions.

CONCLUSION

Macrocrania in infancy due to extra- axial fluid collection in the brain is also referred to as external hydrocephalus or benign enlargement of sub-arachnoid spaces. When familial, it presents with a head circumference greater than the 95th percentile in the parent and has a male preponderance.[5] The aetiology behind extracellular fluid collection in the brain in term infants has been attributed to immaturity of the arachnoid villi.[6] This explains the spontaneous resorption of CSF as the child grows older thereby allowing most children to regain developmental milestones. Kuruvilla et al., reported a decrease in size of subarachnoid spaces 14 months following admission of a 4-month-old infant despite an enlarged head circumference.[7]

Developmental delay in children with BESS and prior history of trauma could be a consequence of damage to the developing brain. Having ruled out history of trauma, infection during pregnancy or other genetic causes like mucopolysaccharidoses or deficiency of vitamin D, our patient was most likely susceptible to developmental delay due to stagnant flow of CSF. Nickel et al., reported gross motor delay in 7 of 9 children who presented with BESS during their first year of life.[6] These children had age-appropriate gross milestones on follow-up, however, 3 children had persistent speech delay. The inability to regain certain milestones can point towards the development of autism spectrum disorder.[4] Thus, BESS may not always have a transient delay in the development of a child bringing to question the benign nature of this condition.[8] The aetiology behind persistent deficit in gross motor and speech domains on follow-up in selective infants with BESS is unknown and further research on this would help clinicians predict prognosis of these infants and arrange for appropriate interventions to prevent long-term deficits.

Characteristic features on MRI include the cortical vein sign which helps distinguish BESS from subdural collections.[9] The cortical veins traverse the enlarged subarachnoid space and lie adjacent to the inner table of the skull in BESS. The cortical vein sign is also visible in cerebral atrophy which can be differentiated from BESS by prominent widening of cortical sulci throughout the brain without enlargement of bifrontal subarachnoid spaces.[10] In a subdural hygroma, collections of subdural fluid cause displacement of subarachnoid space and the cortical veins lie adjacent to the surface of the brain. Additionally, enlargement of subarachnoid space widths are strongly suggestive of BESS. Normal values of the inter-hemispheric fissure, cranio-cortical and sino-cortical widths range

from 6-8.5 mm, 4-10 mm and 2-10 mm, respectively.[8] Our child presented with inter-hemispheric fissure width and sino-cortical widths well above the normal range and a cranio-cortical width close to the upper limit of normal.

Our patient was born at term with normal head circumference. Enlargement of head circumference was observed at about 6 months of age until 11 months, without further increase at follow-up. A possible explanation for the absence of further increase in head circumference at follow-up could be spontaneous resolution of fluid within subarachnoid spaces permitting growth of the brain resulting in a static head circumference. This, in addition to the absence of signs of raised ICP reassured us of the self-resolving nature of the condition and warranted further radiological imaging or surgical intervention unnecessary. Current literature does not indicate shunt placement or medical treatment using acetazolamide in the absence of raised ICP and emphasises on a more conservative approach.[8] Had the child's head circumference continued to increase, primary investigations would focus on measuring the ICP and the management protocol would involve shunting. The role of early shunt placement in avoiding or limiting developmental delay is unclear and further research could help form a clear indication for shunt placement based on clinical presentation, developmental delay and radiological findings.

Benign enlargement of subarachnoid spaces is a self-limiting condition. We would like to encourage further research on the aetiology of developmental delay and to determine whether early shunt placement in children with BESS can minimise the risk of developmental delay.

LIST OF ABBREVIATIONS

Benign enlargement of subarachnoid spaces (BESS), cerebrospinal fluid (CSF), developmental quotient (DQ), magnetic resonance imaging (MRI), computerised tomography (CT), toxoplasmosis, rubella, cytomegalovirus, herpes simplex, and HIV (TORCH), out-patient department (OPD).

DECLARATIONS

Ethics approval and consent to participate: Not applicable

Consent for publication: Appropriate consent has been obtained.

Availability of data and material: The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Authors' contributions: JMR prepared the manuscript, VDS performed critical revision of the manuscript and JY was involved in collection of data. All authors read and approved the final manuscript.

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