SAR Journal of Anatomy and Physiology

Abbreviated Key Title: *SAR J Anat Physiol*
Home page: https://sarpublication.com/journal/sarjap/home
DOI: https://doi.org/10.36346/sarjap.2024.v05i04.004

ISSN 2707-7713 (P)
ISSN 2709-6874 (O)

Case Report

Normal Pressure Hydrocephalus in a Girl Child with Blake's Pouch Cyst

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Article History: | Received: 13.11.2024 | Accepted: 19.12.2024 | Published: 24.12.2024 |

Abstract: A 4 yrs old baby girl presented with history of gait instability with recurrent fall without loss of consciousness since last 1 yr. with recently having urinary and fecal incontinence off and on after being continent earlier. She did not have any febrile illness or convulsion or vomiting. There was no history of consanguinity or birth injury. Examination revealed no facial dysmorphism; her speech and language were delayed with borderline delayed developmental milestones. Her gait was ataxic with hesitant steps, magnetic in nature characteristic of frontal gait disorder. She was provisionally diagnosed as normal pressure hydrocephalus. MRI brain images revealed infracerebellar Blake's pouch cyst with dilatation of lateral, 3rd and 4th ventricles with normal subarachnoid bathing. She was referred to pediatric neurosurgeon for assessment and intervention for possible 3rd ventriculostomy.

Keywords: Normal pressure hydrocephalus (NPH), Hydrocephalus, Gait disturbance, Gait instability, Gait apraxia.

INTRODUCTION

During normal fetal development, the fourth ventricle perforates by 12 weeks of gestation to form the foramen of Magendie, which opens into the subarachnoid space (cisterna magna). If this perforation does not occur, the cerebrospinal fluid produced by the choroid plexus in the fourth ventricle forms a cyst (Blake's pouch), potentially enlarging this space and lifting and rotating the cerebellar vermis. The vermis usually remains intact, but, on occasion, may be compressed.

Normal pressure hydrocephalus is a progressive neurological disorder characterised by the triad of gait ataxia, cognitive impairment and incontinence with presence of ventriculomegaly without any obstruction in CSF circulation or persistently raised intracranial pressure.

Here we present a 4 yrs old girl who showed the features of normal pressure hydrocephalus with her brain MR imaging showing infracerebellar cyst communicating with 4th ventricle, abutting the normally developed vermis, consistent with Blake's pouch cyst. The non-perforated foramen of Mazendie resulted in partiality obstructed hydrocephalus with normal subarachnoid bathing of CSF.

CASE REPORT

A 4 yrs old girl presented with history of recurrent episodes of fall while walking alone for last 1 yr. Her parents denied any history of birth injury or consanguinity.

Her falling was not associated with any loss of consciousness and she would stand up immediately with support from one of her parents. There was no history of any febrile illness or convulsion, headache or vomiting.

Her development milestones were normal from her birth to 1yr and borderline delayed after 1 yr. Her language was delayed with ability to say few intelligible words. She could understand her parents verbal conversation to follow few simple commands. Her movement was restricted to sitting and standing without support and walking a few steps with support. Due to recurrent fall while walking, she would hesitate to take steps alone.

Her parents also complained of her having urinary and fecal incontinence off and on for last 2 months. This started gradually after her being continent earlier about 3.5 yr. age.

On examination her build was sthenic with body weight of 14 kg. and a height of 103 cm. Her head and chest circumferences were within normal range.

There was no laxity in her joints or any weakness in her limbs. There was no facial dysmorphism. Tone of her limbs was reduced, power and deep tendon reflexes were normal. Cerebellar signs could not be examined for her inability to follow commands.

Her gait was hesitant, small strides, magnetic in nature with decreased movement of her ankle and toes characteristic of frontal gait disorder.

With features of ataxia from frontal gait disorder, cognitive impairment and progressive incontinence, she was provisionally diagnosed with normal pressure hydrocephalus (NPH), pending investigations.

Routine investigations of blood, urine and stool did not show any abnormal finding. So was her metabolic profile. EEG sleep record did not show any abnormality. Nerve conduction study of lower limbs was within normal limit. MRI dorsolumbar spine was normal.

MRI brain without contrast showed an infra-cerebellar cyst communicating with 4th ventricle and hydrocephalus from enlarged fourth third, and lateral ventricles with normal subarachnoid bathing.

The cyst location was consistent with congenital persistent Blake's pouch which normally, unlike in the patient, would disappear with formation of foramen of Mazendie during early intrauterine life.

DISCUSSION

During normal fetal development, the fourth ventricle perforates by 12 weeks of gestation to form the foramen of Magendie, which opens into the subarachnoid space (cisterna magna). If this perforation does not occur, the cerebrospinal fluid produced by the choroid plexus in the fourth ventricle forms a noncommunicating cyst (Blake's pouch), potentially enlarging this space and lifting and rotating the cerebellar vermis [1, 2]. The vermis usually remains intact, but, on occasion, may be compressed. Later in intrauterine life the bilateral foramina of Luschka form communication from 4th ventricle to subarachnoid space thus making the hydrocephalus 'communicating', so called normal pressure hydrocephalus (NPH). Occasionally this cyst formed during intrauterine life may be resolved spontaneously and disappear in post-natal life [3].

This child had the triad features of frontal lobe ataxia, incontinence, and cognitive impairment characteristic of normal pressure hydrocephalus with ventriculomegaly and normal subarachnoid bathing.

The MRI Brain

(Sagittal T2- and axial T1- weighted) images of the girl showed enlarged fourth ventricle communicating with a retromedullary, infravermian cystic compartment abutting the normally developed vermis, and hydrocephalus from partial obstruction of ventricular outflow with prominent flow voids in 3rd and 4th ventricles with normal subarachnoid bathing, consistent with Blake's pouch cyst.

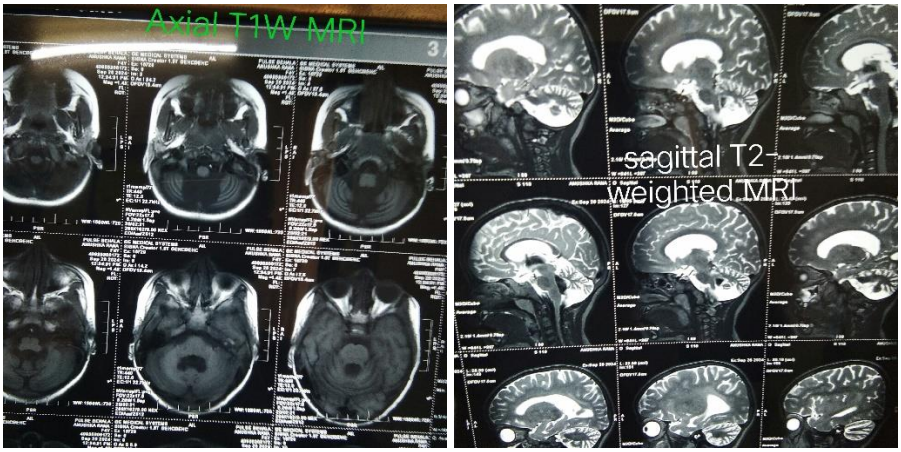

Figure 1and 2: MRI brain images

CONCLUSION

Blakes Pouch Cyst is a posterior fossa cystic malformation that may present with normal pressure hydrocephalus in early childhood.

Cognitive impairment and ataxia do eventually develop. It has characteristic neuroimaging findings that allow to differentiate BPC from other posterior fossa cystic malformations such as Dandy Walker variant, or symptomatic arachnoid cysts. Differentiation between these cystic posterior fossa lesions is important in terms of diagnosis and management.

Conflict of Interest: None

Funding: None

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