Case Report

Indian Journal of Trauma and Emergency Pediatrics
Volume 8 Number 2, May - August 2016
DOI: http://dx.doi.org/10.21088/ijtep.2348.9987.8216.26

Hydrocephalus

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Abstract

Hydrocephalus (from Greek hydro-, meaning "water", and kephalos, meaning "head") is a medical condition in which there is an abnormal accumulation of cerebrospinal fluid (CSF) in the brain. This causes increased intracranial pressure inside the skull and may cause progressive enlargement of the head if it occurs in childhood, potentially causing convulsion, tunnel vision, and mental disability. It was once informally called "Water on the brain"[1].

Keywords: Hydrocephalus; Cerebrospinal Fluid.

Introduction

Hydrocephalus is an abnormal increase in the amount of cerebrospinal fluid within the cranial cavity that is accompanied by expansion of the cerebral ventricles, enlargement of the skull and especially the forehead, and atrophy of the brain.It can be caused by congenital or acquired factors. Congenital causes include Spina Bifida, Arnold–Chiari malformation, craniosynostosis, Dandy–Walker syndrome, and Vein of Galen malformations. Acquired causes include hemorrhage, meningitis, head trauma, tumors, and cysts [2,3].

Case Report

A 12 week old female infant presents to the emergency department with progressive vomiting, lethargy, and difficulty feeding over the past two days. Her mother reports that the infant has been irritable in the last week. She has high pitched cry . She has not been breastfeeding well. The infant has had fewer wet diapers and no bowel movements today. She reports that the infant was born on time and that there were no prenatal or perinatal complications. The infant was discharged after a 48 hour stay in the regular newborn nursery, and had follow-up initially with her pediatrician about 1 week after discharge. She has had no further follow-up. Mother's prenatal labs were normal. The infant weighed 2900 grams at birth (25th percentile), measured 47.8 cm in length (10th to 25th percentile), and had a head circumference of 34 cm (25th percentile).

Examinations

Temperature - 36.5 C, Pulse - 165 beat/min, RR - 45cycle/min, BP - 98/65 mmHg.

Weight 4.20 kg (5th percentile), length 57 cm (10th to 25th percentile), HC 42.6 cm (95th percentile).

In general, this is a lethargic infant with a weak, high-pitched cry. Her head is oddly shaped and looks like an inverted pear. Her scalp veins are prominent, and the anterior fontanelle is tense and bulging. Eyes show pupils which are equal and round, but are sluggishly reactive to light. Red reflex is present bilaterally. Extraoccular movements (EOM) are clearly dysconjugate. She has a downward gaze. There is mild tachypnea with slight intercostal retractions. Lung fields are clear to auscultation bilaterally. Her heart examination reveals tachycardia with a regular rhythm and a systolic ejection murmur at the left sternal border. Capillary refill is 2 seconds. Her spine is straight without protrusions or apparent defects. Her upper extremities show good tone and full range of motion with slightly brisk reflexes. Her lower

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extremities show increased tone with brisk reflexes bilaterally. On neurologic examination, there is a poor suck. The startle response is minimally present. The grasp and glabellar reflexes are present. No parachute reflex can be elicited. The Moro's reflex is present.

Discussion

Pathogenesis

Normal rate of CSF Productionis about 20ml/day.

It occurs because of :

- Increased production of CSF
- Obstruction in flow toCSF
- Decreasaed absorption of CSF.

Types

Communicating Hydrocephalus: Also known as external or nonobstructing hydrocephalus.

It is due to obstruction outside the ventricular system at arachnoid villi or basal cisterns.

Noncommunicating Hydrocephalus: Also called as obstructive hydrocephalus.

The block is at any level in the ventricular system commonly at the level of aqueduct or foramen of Luschka and Magendie[4,5].

Etiology

Congenital Causes

- Intrauterine infections
- Rubella,Toxoplasmosis,cytomegalovirus.
- Congenital malformations:
- Arnold Chiari malformatiion type 1
- Agenesis of Foramin of Monro
- Arteriovenous malformation
- Brain stem malformation
- congenital toxoplasmosis
- Cranial defects
- Dandy Walker syndrome

Acquired Causes

- Infections
  Meningitis
  Mumps
  TORCH infections
  Cysticercosis.
- Post ventricular haemorrhage
- Posterior fossa tumors
- Mass lesions
- Idiopathic.

Pathology

- The ventricles are dilated:ependyma of ventricles may be disrupted.
- Cortex is preserved, but can atrophy with prolonged pressure from the hydrocephalus.
- Cortical atrophy itself may lead to incrased CSF in the ventricular system [6].

Clinical Features

Symptoms

- Headache is more in morning in early stages,becomes better in lying position.
- Vomitting
- Urinary incontinence presenting as urgency, frequency in early stages.
- In late stages there is diminished awareness to need to urinate.
- Blurred vision due to papilloedema.
- Drowsiness
- Horizontal diplopia
- Difficulty in walking

Signs

- Macrocephaly : increase in head circumference of more than 2cm per month in the first 6 months should raise suspicion of hydrocephalus.
- Sutural separation can be palpated
- Fontanels: delayed closure.
- Scalp veins become prominent and dilated.
- Sunset sign
- Macewen sign
- High pitched cry
- Pyramidal signs in the lower limbs: plantor extensor reflex andexaggerated deep tendon reflexes

- in the lower limbs and increased tone in lower limbs are seen.
- Lateral gaze palsy
- Papilloedema
- Bradycardia
- Systemic hypertension
- Altered respiration

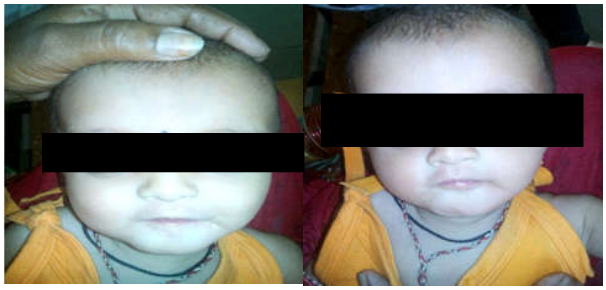

Fig. 1&2: Sunset sign of Hydrocephalus

Investigations

- To find the cause:
  - TORCH screening
  - CSF analysis
  - Ventriculography
  - Pneumoencephalography
- Confirmation of diagnosis

Xray Skull

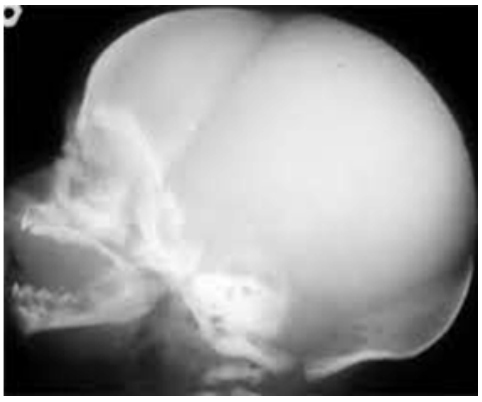

Fig. 3: X ray skull of Hydrocephalus

- Enlargement of skull
- Widening of sutures
- Erosion of clinoid process
- Thinning out of skull bones
- Copper beaten appearance
- Deepening of sella turcica

2. Ultrasonography in infants
3. Ct scan - size of ventricles increased
4. MRI is preferred for posterior fossa and spinal cord lesions
5. Angiography

Treatment

Medical

Temporary relief and incudes use of drugs which decrease CSF secretion or increase CSF reabsorption

- Acetazolamide decreases CSF production it inhibits enzyme carbonic anhydrase dose 50mg/kg/day
- *Loop Diuretics:* Frusemide decreases CSF production]it increases water excretion by inhibiting reabsoption of sodium and chloride in the ascending limp of loop of Henle and distal renal tubule.
- Combination of acetazolamide and frusemide is helpful.

Surgical Treatment[7,8]

Shunt Procedures

- ventriculoperitoneal shunt most commonly used.
- Ventriculoatrial shunt
- lumboperitoneal shunt
- shunt between lateral ventricle and cisterna magna(Tokilsen Shunt)
- ventriculopleural shunt.

Other Surgical Procedures

- Choroid plexectomy
- Choroid plexus coagulation
- Cerebral aqueductoplasty.
- Repeated lumbar puncture
- endoscopic fenestration of floor of ventricle is done in communicating hydrocephalus.

Follow up

- Head circumference shoud be monitored.
- The patients should be taught about the signs of shunt infection or blocks
- Antibiotic prophylaxis should be advised in

patients with vascular shunts

- Periodic re-evaluation should be done
- Patients or care givers should be taught about signs of acute or chronic progressive hydrocephalus.

Prevention

- Early diagnosis and treatment of CNS infections
- Neural tube defects should be prevented by providing folic acid to mother.
- Mother should be screened for intrauterine infections before conception and treated.
- Genetic counselling for genetic disorders.

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