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Case Reports & Case Series

Spontaneous external rupture of hydrocephalus in the occipital region in an infant: A rare case report

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ABSTRACT

With the availability of advanced diagnostic and therapeutic modalities, late presentation of hydrocephalus is a very uncommon occurrence in the present era. Here, we report an 11-months infant with late presenting hydrocephalus with spontaneous rupture at the occipital region.

1. Introduction

Congenital hydrocephalus (HCP) is a condition associated with dilatation of cerebral ventricular systems due to increased accumulation of CSF. The reported incidence of congenital HCP is approximately 0.2–0.5 per 1000 live births [1]. Congenital hydrocephalus may be associated with other anomalies like meningomyelocele or it may occur as an isolated congenital aqueduct stenosis [2,3]. Timely surgical interventions hold the key in having a good neurological outcome. Nowadays, with the availability of ultrasound (USG) and magnetic resonance (MR) screening, prenatal diagnosis had become a norm in developed countries leading to the early institution of treatment [4]. However, sometimes treatment is delayed in resource-poor settings due to ignorance, poverty, social beliefs and serious complications may arise as happened in index case.

2. Case report

An 11-month-old male infant presented with gradually increasing head size since birth. Mother had noticed leaking of fluid from the occipital region. There was a history of fever and refusal to feed since last two days. The child was born at term gestation with an uneventful perinatal period. The prenatal diagnosis of congenital hydrocephalus was made on ultrasound screening during the second trimester. At birth, the head circumference was 41 cm and computed tomography brain revealed ventricular dilatation with a VSI > 40% along with thinned out cortical mantle. There was no other associated congenital malformation. The patient's parents had been advised surgery but they refused and took discharge against medical advice. The child again presented at an age of 1.5-months with gradual enlargement of head size. On the 2nd visit, the head circumference measured 48 cm. But despite counseling and explaining the associated risk of delayed treatment, the parents didn't want admission at that time also. During the next seven months, the parents sought treatment from the local quacks thinking of the disease as a goddess's curse as these practices are still prevalent in some remote parts of our country.

At the time of third visit, the child was febrile (101F), sick, and severely dehydrated. The child was severely malnourished as per WHO classification. The head circumference was measuring 78 cm and sunset sign was present (Fig. 1a and b). On local examination, the sutures were widely separated and multiple decubitus ulcers were seen over the occipital region with CSF leakage (Fig. 2). The head shape was severely distorted with a depressed vertex of the skull due to CSF leakage. In the emergency room, the patient was resuscitated with intravenous fluid and broad-spectrum antibiotics started. After adequate resuscitation, the surgery was planned. But the parents again refused surgery as well as admission and left against medical advice. On follow-up, the child expired at home two days later.

3. Discussion

Spontaneous rupture of hydrocephalus is an extremely rare entity in present era due to improved socioeconomic status, availability of better diagnostic and therapeutic interventions. However, in developing

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Fig. 1. a and **b**: infant with massive enlarged head.

countries; situation is quite different and suitable interventions in a patient with hydrocephalus may be delayed due to religious beliefs and social pressure. Almost all of the cases with spontaneous rupture of HCP are reported from resource-poor countries; among them four are from India (Table 1). External rupture of HCP can be differentiated from fistula as there is slow leaking of CSF through epithelialized tract in later as compared to sudden release with rupture. Long-standing raised intracranial pressure may lead fatal complication like formation of ventricular diverticula, ventriculocisternostomy and herniation of brain, or spontaneous brain and ventricular rupture as in index case. Due to disproportionate growth of head size, the body cannot bear the weight of huge head. As a result, the child prefers to lie in the supine

Fig. 2. Decubitus ulcer at posterior surface of head with CSF leakage.

position for long period leading to pressure necrosis of skin and formation of decubitus ulcers. It also results in CSF accumulation in the dependent part of the brain i.e. the occipital region. Ultimately in untreated cases, due to progressive distension of ventricles, thinning of brain parenchyma and overlying skin necrosis take place and results in spontaneous brain and ventricular rupture. Due to sudden release of intracranial pressure during rupture, severe depression in the vertex of skull is formed giving “alien head deformity” appearance.

Timely intervention is the key to success in congenital hydrocephalus. The goals of successful treatment are to maintain intracranial pressure within a safe zone and minimize the neurological damage. Ventriculo-peritoneal shunt is the treatment of choice in all cases of hydrocephalus. In cases of rupture, the defect needs closure and cranioplasty is required in almost all the cases. The ventriculitis need to be treated with adequate duration of antibiotics. This report also highlights the need of a multi-disciplinary approach in the successful treatment of HCP, which is often lacking in developing countries. The psycho-social counseling of the parents about this potentially treatable anomaly forms an important pillar of treatment in view of long duration of treatment and follow-up. The physiotherapy and rehabilitation exercises should be taught at every visit to prevent permanent neurological sequelae.

4. Conclusion

As most of the cases with late presentations are reported from developing countries, creating awareness in peripheral health workers and educating the parents about the anomaly and importance of early interventions will be helpful in achieving a favorable outcome.

Conflict of interest

None.

Fundings

None.

Table 1
Clinical profile of patients with spontaneous external rupture of hydrocephalus reported in literature.

Study (year)	Country	Age & sex	Prenatal diagnosis	Site of CSF leak	Intervention done	Final outcome
Ezechukwu et al. [5]	Nigeria	3 Y/F	Not available	Right frontal area	None	Expired
Moghtaderi et al. [6]	Iran	8 mn/M	Not Available	Left postparietal area	VP shunt and cranioplasty	Post-operative survived, lost in follow-up
Garg et al. [7]	India	6 mn/F	Not available	Left postparietal area	Not available	Not available
Mishra et al. [8]	India	8 mn/F	Available	Multiple scalp necrosis	Intravenous Antibiotics, planned for surgery	Expired
Tripathi et al. [9]	India	11 mn/M	Not available	Anterior fontanelle	Bone grafting	Survived
Dhyani et al. [10]	India	7 mn/F	Not available	Occipital area	Intravenous Antibiotics, Planned for surgery	Expired
Aggarwal et al. [11]	India	10 Y/M	Not described	Parietal region (Ventriculosubgaleal fistula)	VP shunt	Survived

(M-Male, F-Female, Y-Years, mn-months)

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