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LETTER TO THE EDITOR

Spontaneous rupture: a rare complication of neglected hydrocephalus

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Dear Editor:

There has been improvement in the outcome of treatment of hydrocephalus. This however is not often the case, especially in the developing countries where paucity of skilled personnel, lack of fund, ignorance and religious and social beliefs often present a big obstacle to prompt treatment [5]. In these cases, progression of the condition often results in complications including delayed developmental milestones, cognitive impairment, visual loss, death and very rarely spontaneous rupture [1, 4, 5]. We report a case of spontaneous rupture of neglected hydrocephalus in a Nigerian infant and highlight the underlying socioeconomic factors contributing to this condition in a resource-challenged setting.

A 5-month-old male infant presented with progressive head enlargement since birth and copious leakage of CSF from the right occipital region a day before presentation. Prenatal diagnosis of hydrocephalus was made with transfontanelle ultrasound scan at gestational age of 7 months, and the baby was delivered by elective caesarean section at term, in a rural secondary health facility. He was referred on the 5th day of life to a tertiary hospital with neurosurgical service, but the parents did not go because of lack of fund and ignorance. During the following 5 months, the head size continued to increase until there was an acute spontaneous leakage of CSF observed from the head a day before necessitating the child to be presented to our service.

At presentation, the child was afebrile but dehydrated and malnourished. He was lethargic with weak cry. There was marked craniofacial disproportion and prominent scalp veins. He had upgaze paralysis bilaterally (Fig. 1). There was no light fixation or tracking. The head was disfigured with marked sutural diathesis, scaphoid anterior fontanelle and multiple scalp pressure ulcers (Fig. 2). The head circumference was 73 cm, which was much more than the 97th percentile for age. The site of CSF leak was identified as a 1 cm by 1 cm ulcer at the left occipital region (Fig. 2, arrow). There was no active CSF leak at presentation. Resuscitation was commenced immediately and the child started intravenous fluids and a broad-spectrum antibiotic. He died about 32 h after.

In the developing world like ours, some cases of hydrocephalus are still left untreated [3–5]. In these cases, long-standing tension hydrocephalus can sometimes lead to ventricular diverticula formation or rupture [2, 3, 5]. The ventricle can rupture into the cisterns leading to spontaneous ventriculostomy (ventriculocisternostomy) or through the necrotic scalp resulting in spontaneous brain rupture [3, 5].

In these neglected cases, progressive ventricular enlargement occurs at the expense of thinning of the cerebral mantle, as well as thinning of the skin and subcutaneous tissue. The weight of the head and difficulty with positioning in these patients subsequently result in pressure necrosis of the thinned-out scalp and ulcer formation which ultimately lead to spontaneous rupture of the brain and CSF leakage [3–5].

Spontaneous rupture of hydrocephalus is extremely rare with only few cases reported in the literature thus far [5]. At 5 months, our patient is the youngest case yet. Most of the reported cases are from resource-limited countries like our own where poverty, ignorance, stigmatization and social and religious beliefs contribute to this unwanted condition [5]. Availability and access to a neurosurgeon in these settings can also be a contributing factor particularly due to unequal distribution with most in the major cities, away from the reach of a significant number of citizens.

This present case exemplifies the many challenges in the care of these children in a developing economy despite a

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Fig. 1 A clinical photograph showing macrocephaly, prominent scalp veins, bilateral upgaze palsy and wide sunken anterior fontanelle

prenatal diagnosis and subsequent delivery by caesarean section at a secondary health facility about 52 km from our centre. The distance from the centre to which the child was referred for care, which is about 200 km from the place of residence of the parents, the lack of fund and the misinformation that the child was incapable of living a useful life, peddled by a member of the health team, contributed to the neglect of the care of this child which resulted in a disastrous outcome. This again highlights the impact of poverty, misconception and poor understanding of neurological conditions and their outcomes among the non-specialist health care practitioners in developing countries on the outcome of diseases and health indices in the developing world.

We suggest a further upscale of advocacy to educate the population and even health care providers about this disease condition, availability of effective treatment and possibility of a productive living following treatment. Groups with parents and long-term surviving patients as members will greatly be of help in this regard. Efforts geared towards increasing the numbers of neurosurgeons in these settings should be sustained. Incentives should also be provided to these highly trained specialists to stay at the rural areas.

Fig. 2 A clinical photograph showing the collapsed head with pressure ulcers. The site of the CSF leak was the 1 cm × 1 cm ulcer at the occipital region (arrow)

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Informed consent Informed consent was obtained from all individual participants included in the study.

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