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Case Report

Delayed Death Due to Hydrocephalus - Case Report

Dr. Basappa S Hugar[1]*, Dr. S Harish[2], Dr. Girish Chandra YP[3], Dr. Udaya Shankar BS[4], Dr. Jayanth SH[4]

[1]Assistant Professor, [2]Professor and HOD, , [3]Professor, [4]Post Graduate Student; Dept of Forensic Medicine, MS Ramaiah Medical College, Bangalore-560054.

e-mail id:basavaraj_hugar2007@rediffmail.com

ABSTRACT

Hydrocephalus in children is a condition in which there is excessive accumulation of cerebrospinal fluid (CSF) in and around the brain and spinal cord. Hydrocephalus in most of the circumstances is congenital hence, would be detected early in pregnancy by various available imaging techniques, thus, terminating pregnancy in the initial stage itself and thereby avoiding mental agony to parents. Death due to hydrocephalus is a rare entity especially that produced in the post natal period. One such case of a 18 month old male baby who was subjected for autopsy at Department of Forensic Medicine, M.S.Ramaiah Medical College with alleged history of birth trauma due to wrong forceps application in a private hospital at the time of delivery is being discussed here.

Keywords: Hydrocephalus, Birth injuries, Intra cerebral haemorrhage, Birth asphyxia

INTRODUCTION

It's rare for a Forensic expert to come across in day to day practice the cases due to hydrocephalus. Death due to hydrocephalus as a delayed sequel of birth trauma is rarely encountered in Forensic practice. Hydrocephalus is defined as excessive accumulation of CSF in the ventricles with consequent thinning of the brain tissues and enlargement of the cranium, which occurs 1 in 2000 deliveries[1]. CSF volume is usually between 500ml and 1500ml, but as much as 5L may accumulate. The head circumference often exceeds 50cm and may reach up to 80cm[2]. Hydrocephalus may be congenital or acquired. Congenital hydrocephalus is present at birth, and may be caused by either environmental influence during foetal development or genetic predisposition.

Congenital hydrocephalus is associated with other congenital malformations (aneuploidy) in 1/3rd of the cases[1]. The salient clinical features include large bulging anterior fontanel, separated sutures, bossing of frontal bones, and sun setting sign[3].

Acquired hydrocephalus develops at the time of birth or at some point afterward. This type of hydrocephalus can affect individuals of all ages and may be caused by injury or disease.

Intra cerebral haemorrhage is one of the rare causes of development of hydrocephalus in postnatal period. Intra cerebral haemorrhage can result from various birth injuries.

Birth injuries are important cause of perinatal mortality and morbidity, which are defined as those sustained during the labour and delivery[1]. ICH in the newborn is frequently associated with prolonged or precipitous delivery, vaginal breech delivery, instrumental delivery, use of forceps or ventouse extraction, and primiparity or extreme multiparity[4-7]

Birth injuries most commonly involve head, and the severity can range from minor scalp abrasion to fatal skull fracture. The most common dangers to the foetus involved in forceps application could be immediate for

example, asphyxia, facial bruising, intracranial haemorrhage (rupture of great vein of galen), skull fractures, cervical spine injury and so on ; or remote for example, cerebral or spastic palsy due to residual cerebral injury. Forceps application can result in intra cerebral haemorrhage (Birth trauma). Birth trauma is a frequent accompaniment of birth asphyxia, both being results of difficult and prolonged deliveries[8]. It might be a cause for late development of hydrocephalus and mental retardation. Other causes of intra cerebral haemorrhage are anoxic and primary haemorrhagic disease.

In contrast to the paucity of cases reported in literature, however, intra parenchymal haemorrhage is not uncommon in clinical practice[9]. Setting sign may be normally seen in some neonates, if persistent and exaggerated it suggests hydrocephalus. It is suggestive of raised intracranial tension with compression of orbital plate or brainstem irritation[3]. Hydrocephalus ex-vacuo occurs when there is damage to the brain caused by stroke or traumatic injury. In these cases, there may be actual shrinkage (atrophy or wasting) of brain tissue.

CASE REPORT

A 18 month old male baby was subjected for autopsy on 18 December,2009 with alleged history of birth trauma due to wrong forceps application. On examination% the baby was poorly built for the age, poorly nourished, wheatish in complexion, measured 66 cm in length. Eyes were closed; eyeballs were sunken downwards exposing most of the upper part of sclera (sunset appearance). Post mortem staining was faintly appreciated over back of the body. Rigor mortis was setting in. Head was disproportionately enlarged. Head circumference- 57 cm, chest circumference- 34 cm, abdominal circumference- 25 cm. Teeth were not erupted; scalp hairs measured 10 cm, sparse and grey coloured.

On opening the cranium- bones of vault were thinned out and were not fused with each other (gap of 1 to 1.5 cm). All fontanelle were not fused. Underneath meninges were stretched, upon incision, straw coloured fluid of about 100 ml drained out, cerebrum was thinned out and cerebellum intact. All other organs were intact and pale. Brain, heart, kidneys, liver, spleen was sent for histopathological examination.

Histopathological examination revealed:

Brain – Hydrocephalus (severe degree). Cerebral parenchyma- Signs of compression and atrophy, cerebellum-unremarkable.

Lungs, Kidneys and Spleen – Congested

Heart and Liver – Unremarkable

Further history from the parents about past medical records, the antenatal period was uneventful and the foetus was healthy. On completion of term, the labour was prolonged and a futile attempt to deliver by application of forceps (vacuum and low mid-cavity forceps) was made by giving a right medio-lateral episiotomy. Hence, an emergency LSCS was done and male baby was extracted on next day morning, baby cried after resuscitation. Subsequently, the neonate was kept on ventilator for 8 days and had frequent seizures during the stay and treated. CT scan of the Brain was done which showed Intra ventricular haemorrhage and was diagnosed to be Hypoxic ischaemic encephalopathy (HIE) stage III. The child was discharged once the vitals were stable, but the child was lethargic and gradually the head started increasing in size disproportionately and rest of the body did not develop. All other milestones were not achieved. Meanwhile, the child was taken to various paediatric neurologists/neurosurgeon/paediatric neurosurgeons and others , but everyone expressed their helplessness as the CT and MRI of Brain showed gross irreversible damage and severe increasing hydrocephalus. Subsequently, the child died after $1^{1}/_{2}$ years on18 December,2011.

DISCUSSION

Even though hydrocephalus is most commonly congenital, postnatal development is also a very important one, since it is preventable to the larger extent. Since in the modern era, congenital hydrocephalus are well detected in intrauterine life by means of regular ultrasound abdomen, because of awareness of medical aids during antenatal period. But hydrocephalus developed in the postnatal period as a delayed sequel of birth trauma sustained due to forceps application has to be kept in mind.

Picture No. 1: Head disproportionately enlarged

Picture No. 2: Baby - poorly built for the age and poorly nourished

Picture No. 3: Eyeballs sunken downwards exposing most of the upper part of sclera (sunset appearance)

Picture No. 4: Brain showing thinned out cerebrum and intact cerebellum

The normal birth process itself may be traumatic enough to cause intracranial haemorrhage in term newborns[10]. A retrospective case-control study in 66 term infants imaged within 7 days after birth showed an increased risk of intracranial haemorrhage with forceps-assisted delivery[11].

In a study they concluded that if attempts at vaginal delivery fail, the risk of injury are increased no matter which method of delivery is chosen[12]. The important conclusion from the study by Towner et al.[13] was that successful vaginal delivery with the use of either vacuum extraction or forceps appeared to carry no excess risk of intracranial haemorrhage, compared with caesarean section during labour.

In the present case, the doctors claimed that the development of hydrocephalus was due to severe birth asphyxia and HIE which might have led to the intra ventricular haemorrhage and subsequent development of hydrocephalus. But relatives alleged that wrong application of forceps has led to the development of intra ventricular haemorrhage and delay in the decision to take for an emergency LSCS after prolonged efforts by forceps which is the prime cause for birth asphyxia and HIE.

After taking into consideration autopsy findings, HPE report and procuring the hospital case records and brief facts of the case, the cause of death was given as “Death is due to raised intracranial tension due to hydrocephalus.”

CONCLUSION

Since the development of hydrocephalus could not be attributed exactly either to the HIE/Birth asphyxia or to the IVH due to forceps induced birth trauma as hydrocephalus is a delayed sequel of both HIE and the IVH due to forceps induced birth trauma. Hence, the cause of death was given as “Death is due to cardio respiratory failure as a result of raised intracranial tension due to hydrocephalus.”

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