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Spontaneous rupture of congenital hydrocephalous: a rare complication

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COMMENTARY LETTER

Familiar periventricular nodular heterotopia in a 1year old female child

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Abstract:
Hydrocephalous represents a diverse group of conditions that result from impaired circulation and/or absorption of CSF, or in rare circumstances, from increased production of CSF by choroid plexus papilloma. Hydrocephalous could be classified as communicating and non communicating. Hydrocephalous resulting from obstruction within ventricular system is called obstructive or non communicating hydrocephalous whereas that resulting from obliteration of subarachanoid cisterns or malfunction of arachanoid villi is called communicating or non obstructive hydrocephalous. Both forms can be congenital or acquired. The first surgical treatment reported in literature was performed by Hippocrates in 5th century B.C., who punctured the lateral ventricle in a patient with obstructive HC. Several procedures and devices have been developed since then, such as external ventricular drainage, external lumbar drainage, repeated lumbar puncture, Ommaya reservoir insertion, or ventriculosubgaleal diversion, ventriculoperitoneal and ventriculoatrial shunts. In untreated cases, head size may continue to increase. Spontaneous external rupture of hydrocephalous is rare presentation. Very few cases have been reported till date.

Keywords: *hydrocephalous, external spontaneous rupture*
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Sirs,

Hydrocephalous represents a diverse group of conditions that result from impaired circulation and/or absorption of CSF, or in rare circumstances, from increased production of CSF by choroid plexus papilloma (1). Hydrocephalous could be classified as communicating and non communicating. Hydrocephalous resulting from obstruction within ventricular system is called obstructive or non communicating hydrocephalous whereas that resulting from obliteration of subarachanoid cisterns or malfunction of arachanoid villi is called communicating or non obstructive hydrocephalous. Both forms can be congenital or acquired (2). The first surgical treatment reported in literature was performed by Hippocrates in 5th century B.C., who punctured the lateral ventricle in a patient with obstructive HC (3). Several procedures and devices have been developed since then, such as external ventricular drainage, external lumbar drainage, repeated lumbar puncture, Ommaya reservoir insertion, or ventriculosubgaleal diversion, ventriculoperitoneal and ventriculoatrial shunts (4).

Clinical features of hydrocephalous will depend upon age of onset, nature of lesion and duration and rate of increase of intracranial pressure. In infants, these include increased rate of head growth, wide open bulging fontanelles, dialated scalp veins, irritability, vomiting, poor feeding, setting-sun eye sign, spasticity, brisk tendon reflex, clonus and babinski sign. In neglected cases, head size may continue to increase. Spontaneous external rupture of hydrocephalous is rare presentation. Very few cases have been reported till date (5-8).

Figure 1. Ruptured hydrocephalous with overlapping of skull bones and sun setting eyes.

A 7 month old female child, born at term out of non consangious marriage through normal vaginal delivery at home. The child presented to our institution with complaints of progressive increase in head size since birth with vomiting since last 4 days and leaking of fluid from occiput with rapid reduction of head size with collapse of skull bones since 2 days. Parents had noticed unusually large head size when child was around of 15 days of age. CT of head done at that time showed Aqueductal stenosis. Surgery was advised but was not done due to financial reasons. On examination, there was overlapping of skull bones and CSF leak was present at occiput (Figure 1). The child was temperature of 97.6 F, heart rate of 168/minute with feeble pulses, respiratory rate was 46/minute There was hypotonia with grade 2 power in all 4 limbs. Complete blood count showed hemoglobin of 9.8 g, leucocyte count of 18,200/mm^3 with 84% polymorphs. There was no improvement after fluid resuscitation and vasopressors were added. Broad spectrum antibiotics were added but the child succumbed within 4 hours of hospital admission.

In hydrocephalus, increased pressure in ventricular system leads to enlargement of head size. This enlargement occurs at the expense of cortical mantle initially and hence cortical thinning is a common associated finding. In neglected cases, further increase occurs at the expense of skin and subcutaneous tissue. Due to weight of the oversized head, infant is unable to hold his neck and is kept supine for most part of the day, pressure necrosis of skin of occiput increased risk of external rupture of already compromised area. Spontaneous rupture is very unusual presentation. Amongst the 4 previously reported cases three are from India (5-8). Another interesting point to note is that all except one of those reported were females, due to cultural neglect of female gender.

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