External Hydrocephalus: A Report of 16 Cases from Oman

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Summary

Sixteen cases of external hydrocephalus (EH) were seen from January 1993 to June 1995. There were 13 (81 per cent) male and three female children. Fourteen (88 per cent) were under 12 months of age. Three siblings with EH were seen in one family. All, but three recovered over time without medical or surgical intervention. These three needed cerebral decongestants in the acute phase. This is the first report of EH from Oman.

Introduction

EH is a condition characterized by increased collection of cerebrospinal fluid (CSF) in the subarachnoid space, predominantly bifrontal, with widened interhemispheric fissure and minimal or no enlargement of the ventricular system as seen on CT scan of the brain.1 A recent rapid increase in head circumference, with above criteria are diagnostic of EH.2 3 No definite aetiology is known. Infection, trauma and metabolic factors have been postulated.3 4 Familial cases have also been reported.5 The condition is usually self limiting with no morbidity.2 3 This condition has to be differentiated from cortical atrophy and subdural effusion.

Materials and Methods

All children with recent rapid increase in head size and varying associated complaints referred to the Paediatric Neurology division of Sultan Qaboos University Hospital from January 1993 to June 1995, formed the subjects of the study. A detailed history regarding development, head trauma, drug ingestion, infections, vomiting, and excessive irritability were recorded. All these children had detailed neurological assessment. The head circumference (HC), height and weight were plotted in growth charts. The national child health services of Oman provide WHO health cards (mandatory for each child from birth to 15 years) indicating all the events in the child's development, including all illnesses. The children with HC >97th centile and normal height, weight (Fig. 1) and development were clinically suspected to have EH. Besides full blood count, routine biochemistry, and relevant metabolic work up, computed tomography (CT) scan head was used to diagnose the condition (EH), and differentiate it from different causes of macrocephaly. The diagnostic criteria was based on CT scan features devised by Maytal et al.1 These criteria being: (i) widened subarachnoid space in the frontal region on both sides with normal or near normal subarachnoid spaces in other areas; (ii) no or mild ventricular enlargement; and (iii) widening of interhemispheric fissure extending posteriorly to middle third of brain. Magnetic resonance imaging (MR) was not done in any child as it was not available in the country. The CT scan pictures were graded depending on the degree of widening of subarachnoid spaces in the bifrontal and interhemispheric fissure. A width of 2–3 mm was labelled as grade I, 3–4 mm grade II, and more than 4 mm as grade III. The CT scans with features of cortical atrophy uniform increase of entire subarachnoid space with ventricular enlargement, and normal or below normal HC, subdural effusion (asymmetric or symmetric enlargement of subarachnoid space with pressure effect on cerebral cortex) and other causes of macrocephaly were excluded. Cerebrospinal fluid (CSF) examination was done in the first four cases, which were normal. Subsequently, CSF examination was done if indicated. EEG was done only in three children. All the children with EH were followed up at Paediatric Neurology out-patient clinic over the last 3 1/2 years.

Results

Sixteen children with EH were identified of which 13 were male and three were female. Fourteen (86 per cent) were below 1 year (Table 1). The mean age was 7.9 months. All children were referred with recent (1–2 weeks) rapid increase in HC. Anterior fontanelles were wide open, pulsatile, bulging in four cases (25 per cent). Associated features included mild upper respiratory tract infection (n = 5; 31 per cent), irritability (n = 5; 31 per cent), vomiting (n = 4; 25 per cent), fever (n = 1), ataxia (n = 1), and seizures (n = 1). These features were seen together or in isolation. The development of motor milestones, language and social skills were normal in all. There were 3 male siblings in one family seen over 3 years. All these presented in age group less than 1 year.
Systemic and neurological examination were normal in all except one. This child had cerebellar ataxia. Papilloedema was not seen in any child. The HC was above 97th centile at presentation in all the cases. Normal HC was noted at birth and 1-2 weeks before the presentation in all cases. Other anthropometric parameters like weight and height remained normal for their age (Fig. 1). The typical CT scan features were as seen in (Fig. 2). The CT scan showed grade I EH in eight children, grade II in four, and grade III in the remaining four. EEG done was normal in two and the third showed nonspecific slowing. Cerebrospinal fluid was normal in all four (initial four cases). The blood counts, electrolytes, liver and renal function were essentially normal in all. One child with cerebellar ataxia and fits was found to have organic acidemia. The children with vomiting, irritability and bulging fontanelle were given two or three doses of intravenous 20 per cent mannitol in the first 24 h and later followed by oral acetazolamide for 2-3 weeks.

On follow-up the HC plateaued over several months and then became normal for age again after about one year. The CT scans were repeated after 1-2 years in six cases (two familial cases). All the features of EH on CT scan had completely disappeared (Fig. 3).

**Table 1**

<table>
<thead>
<tr>
<th>Age (months)</th>
<th>Male</th>
<th>Female</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-6</td>
<td>5</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>6-12</td>
<td>6</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td>12-24</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>&gt;24</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>13</td>
<td>3</td>
<td>16</td>
</tr>
</tbody>
</table>

Discussion

Non-traumatic pericerebral collections of fluid (external hydrocephalus) represent one of the common causes of large head size in infants and children. This is not widely recognized. Since the widespread use of latest neuro-imaging techniques, cases are increasingly reported from different countries. This is the first report from Middle-East as Med-line CDROM search of the last 10 years did not reveal any study. EH can be symptomatic or asymptomatic. The latter condition is also labelled as benign EH. The frequency of symptomatic EH has been reported as 14 to 57 per cent. The exact mechanism for production of EH is controversial. Impairment of CSF resorption due to increased resistance in arachnoid villi or increased venous pressure favours subarachnoid collection.
while collection in subdural space is explained, on subarachnoid subdural fistula with one-way valve effect or through other undefined postulated mechanisms. The exact location of fluid in subarachnoid space or subdural space is believed to be difficult on CT scan, though contrast CT may help in few cases. In a recent study, MRI was found specific to differentiate the benign enlargement of subarachnoid spaces from subdural collections. A given patient may have combined fluid collection in subarachnoid space as well as subdural space, well demarcated by MRI. The usual age group for presentation has been in the first year of life, the main symptom being recently noted big head. Most of our cases presented under one year of age, the majority being in the 3–9-month age group with recent out of proportion head enlargement as compared to body weight and height/length (Fig. 1). This has been noted in an earlier study in 72 per cent of the cases. Thirteen out of 16 cases were males in our study. The male preponderance has been seen in an earlier study. EH patients are
usually asymptomatic, but evidence of intracranial hypertension has been found in 15 per cent of cases in a study.11

Four patients (25 per cent) which presented with vomiting, irritability, and bulging fontanelle responded very well to mannitol and acetazolamide. Thus, we believe EH is associated with raised intracranial pressure in a proportion of cases and a few of them may have raised pressure for a long time needing prolonged cerebral decongestants. There were no definite causes found in the aetiopathogenesis of benign EH. Five cases had features suggestive of viral infections (self-limiting febrile illnesses involving upper respiratory tract). One of our cases had organic acidemia. Association of a child with glutaric aciduria type I has been reported.4 This association seems to be coincidental. Familial forms and identical twins having EH have also been reported.11,13 In our study, three children in one family, all males, had EH. This may suggest benign X-linked or autosomal recessive inheritance, not reported before.

All children had normal neurological development and continued to do so on follow-up, although there are reports of mild developmental delay in some of these children.11 The CT scans repeated in six cases showed complete disappearance of the features of EH (Fig. 3). The HC was normal by 24–36 months of age in all.

The diagnosis of EH should exclude known causes like subdural haematoma and effusion, genetic syndromes, and metabolic diseases. The course of the majority of cases is benign, although surgical treatment has sometimes been performed.14 This is an age-related self-limiting condition occurring in infants, that usually resolves without any active intervention by 2–3 years of age.

Conclusions

EH is a benign condition which is seen mainly in infancy when the anterior fontanelle is open and most of the major cranial suture are not fused. No causative factors of significance were seen, though mild infections were noted in a few cases at the onset. This is a transient phenomenon of unbalanced CSF dynamics which is self-limiting and, hence, does not usually require any medical or surgical intervention. We hypothesize that EH is a condition which evolves out of a transient imbalance between CSF production and absorption by arachnoid granulations, in most of the cases.

Autosomal recessive forms of EH have also been reported. Our study included three sibs from the same family, which suggests a genetic basis for susceptibility to EH in a few.

References