Clinical Reports

Cranial diabetes insipidus secondary to arrested hydrocephalus

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Summary: An unusual case is described linking cranial diabetes insipidus with longstanding arrested hydrocephalus. The latter was demonstrated by computed tomographic (CT) and nuclear magnetic resonance (NMR) scans and cerebrospinal fluid pressure measurements. The increasing use of CT and NMR scans may result in this association of cranial diabetes and hydrocephalus being better defined.

Introduction

Cranial diabetes insipidus (CDI) is a syndrome which has many causes. Hydrocephalus is a rare but recognized cause.¹ The onset of CDI in non-neoplastic and non-traumatic cases is usually within the first two decades.² We report a case of CDI secondary to longstanding hydrocephalus presenting late in life.

Case report

A 48 year old computer programmer was admitted with a 4-week history of an exacerbation of colonic Crohn’s disease diagnosed by barium enema and rectal biopsy two years previously. Systems review revealed a 30-year history of polydipsia and polyuria. The patient was taking no medications. Past medical history revealed no tuberculosis, meningitis or head trauma. The last menstrual period was 2 months prior to her present admission. She had one child, a son, aged 26 years.

Physical examination was unremarkable and in particular the fundi were normal. Investigation showed a serum sodium ranging from 150-160 mmol/l, rising to a maximum of 175 mmol/l 4 days following admission. Potassium, bicarbonate and urea remained normal as did the serum protein and calcium levels. A random glucose was 6.3 mmol/l and plasma thyroxine was 83 nmol/l. A tine test was positive and a Kveim test was negative. A lateral skull X-ray was normal and the Goldmann field of vision test showed no deficit. Computed axial tomography (CT) scanning of the brain showed dilatation of the third and lateral ventricles but the fourth ventricle appeared normal (Figure 1a and 1b). A nuclear magnetic resonance (NMR) scan confirmed communicating hydrocephalus (Figure 2a and 2b). Intracranial pressure was measured throughout a 24-hour period via a right frontal reservoir and as the pressure did not rise above 10 mmHg, a diagnosis of arrested hydrocephalus was made.

A standard water deprivation test revealed a failure to concentrate the urine until intramuscular desmopressin (DDAVP) in a dose of 10 μg was given, suggesting a diagnosis of CDI (Table I). In order to further assess pituitary function a combined insulin stress, thyrotrophin releasing hormone (TRH) and luteinizing hormone releasing hormone (LHRH) test was done. After 8 units i.v. of soluble insulin, the patient became clinically hypoglycaemic at 30 minutes with a fall in plasma glucose from 4.2 to 0.6 mmol/l. The plasma growth hormone response was normal with levels at 0, 30 and 60 minutes of 5.4, 68.6 and 24.4 mU/l, but...
there was some impairment of cortisol response with levels at 0, 30, 60 and 90 minutes of 190, 234, 329 and 252 mmol/l. The thyroid stimulating hormone (TSH) response to 200 μg of i.v. TRH was normal with 0, 20 and 60 minute TSH values of 2.1, 9.1 and 8.2 mU/l. The follicle stimulating hormone (FSH) and luteinizing hormone (LH) response to 100 μg LHRH showed a post-menopausal pattern with values for FSH at 0, 30, 60 and 90 minutes of 3.5, 7.3, 11.7 and 12.2 mU/l and the corresponding results for LH were 11.3, 49.7, 75.7 and 83.7 mU/l respectively.

A diagnosis was made of late presentation of CDI secondary to longstanding hydrocephalus with impaired hypothalamic-pituitary function. Treatment with intranasal desmopressin in a dose of 10 μg twice daily resulted in a resolution of her polydipsia and polyuria and the serum electrolytes became normal. She was given hydrocortisone 10 mg per day to correct the partial hypoadrenalism.

Discussion

Cranial diabetes insipidus is a clinical syndrome of thirst and polyuria due to a failure of adequate quantities of vasopressin to be released into the circulation despite adequate osmotic stimulus. CDI is idiopathic in one third to one half of cases, usually sporadic but may be autosomal dominant. Recent work has suggested an autoimmune aetiology in some cases of the idiopathic group.

Figure 1 (a) CT scan showing dilatation of the third and lateral ventricles. (b) CT scan shows a normal fourth ventricle.

Figure 2 (a) Coronal magnetic resonance scan revealing enlargement of the third and lateral ventricle. (b) Sagittal magnetic resonance scan confirming a normal-sized fourth ventricle.
Table I  Results of the 6-hour water deprivation test and subsequent response to 10μg of desmopressin given intramuscularly

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<thead>
<tr>
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<th>Osmolality (mmol/kg)</th>
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<tbody>
<tr>
<td></td>
<td>Plasma</td>
</tr>
<tr>
<td>Before deprivation</td>
<td>287  97</td>
</tr>
<tr>
<td>6h after deprivation</td>
<td>296  162</td>
</tr>
<tr>
<td>Urine osmolality ratio</td>
<td>at 6 h = 0.54 (normal range &gt; 1.9)</td>
</tr>
<tr>
<td>2h response to DDAVP</td>
<td>308  375</td>
</tr>
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The identifiable causes of CDI are many and clearly listed elsewhere.1 2 3 A modern textbook of neurology recognizes the rare association between hydrocephalus and CDI but no references are cited.1 A major review of the aetiology of CDI in 92 cases identified no cases of hydrocephalus.2 Another review of cases of CDI presenting in childhood identified intracranial defects in 14% of cases.5 Hydrocephalus is noted as a very rare association in this series but no figure is given for its incidence. The changing pattern of the aetiologies of CDI coupled with a reducing incidence of the idiopathic group is consistent with our improving diagnostic abilities.5 Therefore some cases which would have been labelled as idiopathic in the days before CT scans may have been due to intracranial defects.

In CDI a lesion of the neurohypophysis or paraventricular nuclei of the hypothalamus is present. We can postulate that hydrocephalus can lead to CDI by distension of the third ventricle and compression of the above named structures. This theory is given weight by the evidence that hydrocephalus is known to cause obesity and genital hypoplasia by a similar mechanism.1

This case is rare and shows clearly the association of CDI with hydrocephalus in an adult. In addition there was evidence of impaired cortisol response to stress and it was possible that there was a deficit of corticotrophin releasing factor as well.

References


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