Etiologies of Central Diabetes Insipidus in Children

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Abstract

Background Central diabetes insipidus is a heterogeneous disease mainly due to lesions of hypothalamic nuclei or pituitary stalk caused by tumors of the suprasellar region or infiltrative diseases such as Langerhans cells histiocytosis or sarcoidosis.

Objective: The aim of our study was to analyze the etiologies of central diabetes insipidus in children.

Methods: Medical records of 31 children (mean age of 7 years) were reviewed.

Results Craniopharyngioma was the main etiology found in 71%, in 3.2% diabetes insipidus was secondary to autoimmune hypophysitis, in 3.2% it appears after radiotherapy for a germinoma of the pineal region, finally, in 22.6% it was considered as idiopathic.

Keywords Diabetes insipidus; Craniopharyngioma; Hypophysitis; Germinoma

Introduction Central diabetes insipidus (DI) is a rare disorder in children. It can result from a multitude of etiologies including tumors, malformations, traumatic processes, autoimmune, infiltrative diseases, mutations in the gene of arginine vasopressin and finally idiopathic DI which is a diagnosis of exclusion. The aim of our study is to analyze the different etiologies of central DI in children in our department.

1 Materials and Methods

It is a retrospective study including 31 children presenting central DI. Diagnosis of central DI was based on the presence of polyuria and polydipsia, urine specific gravity of less than 1005 and increase in urine specific gravity in response to Desmopressin. All patients underwent brain imaging study either MRI and/or CT scan. Other diagnosis workup included anterior pituitary function, skeletal survey, determination of β hCG and α foetoprotein in cerebrospinal fluid (CSF) and serum and CSF cytology in case of suspicion of germinoma.

2 Results

Etiology of central DI was as follows, craniopharyngioma in 71% (N=22), idiopathic in 22.6% (N=7), Germinoma in 3.2% (N=1) and autoimmune in 3.2% (N=1).

Among the 22 craniopharyngiomas, in 22.7% diabetes insipidus was present preoperatively and in 72.3% it appeared postoperatively. We compared craniopharyngiomas between patients presenting DI preoperatively and those presenting DI postoperatively (Table 1). We also compared patients presenting DI after craniopharyngiomas’ surgery with a group of patients operated on for craniopharyngiomas and not complicated by DI (Table 2).

| Table 1 Comparison between DI appearing before or after craniopharyngioma surgery |
|-----------------------------------------------|
| Mean Age                        | DI Preoperatively | DI Postoperatively | p   |
|-----------------------------------------------|
| Pituitary deficiency: Global-Partial-Absence | 6.5 years         | 8.5 years          | 0.18|
| Tumor Diameter (MRI)                | 43.4mm            | 38.6mm             | 0.19|
| Tumor Composition: Cystic – Cystic and Solid - Presence of calcifications | 60% - 40% - 60%   | 64.7% - 35.3% - 35.3% | 0.56|
Eight of our patients presented pituitary stalk thickening, among them one had germinoma confirmed by stereotaxic biopsy. One had autoimmune DI due to the association of other autoimmune diseases. Six patients were deemed to have idiopathic DI due to the fact that DI was isolated along with the absence of elements in favour of germinoma, infiltrative disease or autoimmune disease. In addition after a long period of follow up pituitary stalk thickening either disappeared or was stable and no arguments for a specific disease appeared.

### Discussion

In our study craniopharyngioma was the more prevalent etiology of central DI. DI appeared mostly in the postoperative period. Although patients presenting DI preoperatively seems to be younger and had larger tumors than those presenting DI postoperatively, it didn’t reach statistical significance. We didn’t find other predictive factors for the presence of DI preoperatively. The extent of tumor resection predicts the occurrence of DI postoperatively. DI is more frequent after complete tumor resection as reported by other authors (Choux, 1991; Crowley et al., 2010).

Isolated pituitary stalk thickening poses a diagnosis challenge. In children germinoma is the first etiology to think about, this diagnosis should be raised in the presence of anterior pituitary deficiencies, in case of progression of the lesion or if CSF markers are positive, a biopsy is warranted in these cases (Mootha et al., 1997). Other etiologies considered in the evaluation of a pituitary stalk enlargement in children are Langerhans’ histiocytosis and autoimmune DI (Scherbaum et al., 1985). In the absence of other findings DI is called idiopathic, this may be a form of infundibuloneurohypophysitis (Maghnie et al., 2000). However, this may be a diagnosis of exclusion and patients should have close follow up with serial MRI imaging, in idiopathic DI pituitary stalk thickening either resolves or shows no progression.

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