CASE REPORT

A Rare Cause of Diabetes Insipidus in Children: Langerhansian Histiocytosis

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ABSTRACT
Diabetes insipidus is a disease in which large volumes of dilute urine (polyuria) are excreted. We report the observation of a 2-year-old child who presents with stagnation in weight with polyuria polydipsia of diabetes insipidus, the cause of which is Langerhansian histiocytosis. Through this work, we highlight the clinical, paraclinical, evolutionary, and therapeutic aspects of this affection.

KEYWORDS: Polyuria, Polydipsia, Langerhansian histiocytosis.

INTRODUCTION
Diabetes insipidus (DI) is defined by an inability to retain free water and is due either to an insufficient release of antidiuretic hormone (ADH, also called vasopressin), by the hypothalamus (= central diabetes insipidus - DIC), or to a resistance of the kidney to DHA (= nephrogenic diabetes insipidus), clinically diabetes insipidus is manifested by polyuria polydipsia. It is a rare disease with a prevalence of about 1/25,000 cases. DIC is the most common form of diabetes insipidus.

We report an observation of a 2-year-old child who consults for polydipsia polyuria, for whom the investigations revealed Langerhans’-cell histiocytosis (HL).

CASE REPORT
This is a 2-year-old boy, with no particular history, including a good birth weight, healthy until 4 months ago when parents note a stagnation with extreme thirst reaching up to 5 liters per day. The admission examination finds an apathetic child at 2DS for weight with signs of denutrition (hair loss and dander, reduced brachial circumference) in addition to signs of dehydration; Abdominal distension is noted without organomegaly, cutaneous involvement or abnormality of the urinary strip.

An initial assessment reveals no biological abnormalities, in particular no hydroelectrolytic disorders.

After collecting urine, the child was polyuric at 500 ml/kg/day with a low urinary osmolarity at 180 mosm/l. In front of this polyuria, a renal ultrasound is asked back in favor of a hypotonia of the excretory cavities, the balance is completed by an encephalic MRI and CT in favor of lytic bone lesions of the bones of the skull and the face with an infiltration of the post-hypophysis (Figure 1,2,3,4et 5).

In order to compare the biological and radiological data, a bone biopsy is performed whose anatomopathological study and immunohistochemical study is in favor of Langerhans’-cell histiocytosis. The extension assessment is normal, having concluded a low grade histiocytosis and the patient is put under therapeutic protocol in association with desmopressin nasally.
DISCUSSION

Usually, after the discovery of a polyuria-polydipsia it is first necessary to eliminate diabetes through a urine test strip, then to measure the urinary osmolarity and propose a restriction test to distinguish between the DIC and the nephrogenic ID.

Among the organic causes of central diabetes insipidus in children, tumors and cerebral malformations are the most common. Langerhansian histiocytosis is responsible for less than 10% [1,2]. Localizations in the hypothalamic, pituitary and/or posthypophysis regions explain the deficit in antidiuretic hormone (ADH). Diabetes insipidus is usually noted at the time of diagnosis, rarely during relapses [1,2].

The diagnosis of lesions is made by MRI, which reveals a tumor infiltrate in the hypothalamic region, a thickening of the stem, and/or a loss of the signal of the posthypophysis as in the case of our patient [3]. The association between HL and diabetes insipidus is described as the first description of the disease by Hand in 1893. The proportion of patients whose evolution is complicated by pituitary involvement varies between 20 and 25% in a large series of patients. [4,5].

The disease takes many forms. It can reach all organs of the body, the most frequent localization is bone, followed by skin then other lesions: lung, marrow, nervous system. The endocrine localizations, as in the case of our patient, are localized, so the presence of polyuria-polydipsia syndrome suggests hypothalamic-pituitary involvement.

Overall, most localizations are unique (those of bone and skin in particular). In some forms, the disease affects several organs. They are said to be multisystemic, thus posing the problem of treatment and prognosis [2-5].

The diagnosis of HL is carried out by histological analysis demonstrated the characteristic proliferation of Langerhans cells (CL). In light microscopy, CL are recognized by their bypassed nucleus, their pale cytoplasm, weakly eosinophilic containing little or no particles phagocytic. The Langerhansian nature of these cells must be confirmed by the immunohistochemistry of the CD1a membrane antigen or by the presence of Birbeck granules in electron microscopy, whereas the expression of the S100 protein, which has been widely used for a long time, is not specific at present [6].

After having made the diagnosis, it is important to make an extensive assessment of this disease to decide on the therapeutic approach. This assessment has several stages: complete clinical examination, skeletal and pulmonary x-rays, bone marrow study, abdominal ultrasonography and is continued according to clinical signs and abnormalities of the first-line assessment.

The treatment of Langerhans' histiocytosis remains controversial at present and is either localized in cutaneous forms or the involvement of the external auditory canal with Chlormethine [3,7].

The use of surgery (curettage, small resection) is done in localized bone forms.

In the severe multifocal forms, many drugs are used: non-steroidal anti-inflammatory drugs (NSAIDs), steroids, cytostatic drugs such as vincristine, vinblastine, VP-16.
(etoposide), 6-mercaptopurine, methotrexate, cytarabine, and cladribine (2 Cda).

To this list, we can add many drugs considered immunosuppressive or immunomodulating agents: alpha interferon, anti-TNF-α, ciclosporin A, thalidomide and various other drugs such as imatinib, retinoids, and bisphosphonates [3].

The prognosis of this affection depends on the classification of Lahaye-Osband. Thus, the age, the number of initial locations, organ dysfunction and the initial response to treatment are the four elements on which the prognosis depends. [8]

Through our observation, we emphasize the need to explore the pituitary hypothalamic region by imaging in front of polyuria polydipsia, this to eliminate an expansive process including histiocytosis whose prognosis in localized forms is completely different than that of multifocal disorders.

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REFERENCES

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