

Search Histiocytosis X facing insipid diabetes with thickened pituitary stalk

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INTRODUCTION

Insipid diabetes is a **rare disease** in pediatric endocrinology.

Facing a thickened pituitary stalk on MRI pituitary, the main diagnosis to mention are: dysgerminoma, histiocytosis, sarcoidosis, autoimmune hypophysitis [1].

Central Diabetes Insipidus can be the first manifestation of Langerhans Cell Histiocytosis [2].

Histiocytosis is a rare and often **underdiagnosed cause**.

CASE REPORT

We report the case of a teenage girl who presented polyuria - polydipsia syndrome at the age of 14 years consequent full central insipid diabetes confirmed by water restriction test.

MRI showed **pituitary stalk thickened** measured at 2.9 mm with a **loss signal of post-pituitary**.

The initial analysis were negative (blood and cerebrospinal fluid markers of dysgerminoma, body skeletal radiographies, bone scintigraphy, ear scan, lung scan, auto-immune research).

The patient was treated with Desmopressin.

Later, the pituitary stalk has grown to 5 mm (**Figure 1 and 2**) with apparition of an asthenia, a weight gain, and **several anterior pituitary deficits** in gonadal and somatotrophic axis. The Growth Hormone Deficit (GHD) could not be supplemented until the diagnosis of dysgerminoma had not been dismissed.

A stalk biopsy was performed at the age of 15 1/2 showing nonspecific inflammatory tissue and **CD1a + marker** in favour of **Histiocytosis X**. GH was started after the diagnosis confirmation.

Complementary analysis showed bone defects (**Figure 3**) and asymptomatic atypical lung nodules which were not present during the initial radiographies.

After two years, the last MRI showed a pituitary stalk increased to 9 mm with compression on the left side of optic chiasm (**Figure 4**), posing an indication of systemic treatment for Histiocytosis.

CONCLUSION

Histiocytosis X is a difficult and late diagnosis. **Annual repetitions** skeletal radiographies in search of bone's lesion, skin biopsy, lung scanner [2] and repeat pituitary MRI before placing the indication of tige biopsy must be part of the key elements of diagnosis.

References

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Figure 1 : Sagittal Pituitary MRI



Figure 2 : Coronal Pituitary MRI

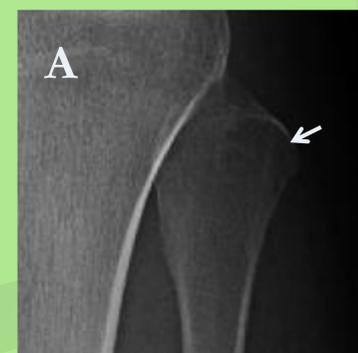


Figure 3 : Fibula (A) and mastoid (B) bone defects



Figure 4 : Coronal Pituitary MRI Compression of optic chiasm