LANGERHANS CELL HISTIOCYTOSIS WİTH ISOLATED CENTRAL DIABETES INSIPIDUS, LOW GRADE FEVER AND CELLAR EROSION

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INTRODUCTION

The annual incidence of Langerhans cell histiocytosis (LCH) is 5 per million in admission with a diagnosis of isolated central diabetes insipidus (CDI) in children under the age of 15 (1).

AIM

The process leading to the diagnosis of LCH at presentation with isolated CDI, imaging findings, and the sellar erosion, will be discussed.

METHOD

Case presentation:

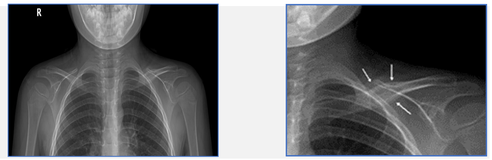
- A 4-year-5-month-old male patient was referred to our outpatient clinic with complaints of drinking too much water and urinating frequently for 2 months. Physical examination, complete blood count, and biochemical tests were normal
- Other pituitary functions were found to be normal. In the follow-up of the patient, it was observed that his body temperature rised to 38-38.4°C in the evening once a day, and fell spontaneously and did not persist. On physical examination, any focus to explain the fever was not found. In laboratory tests, increases in acute phase reactants (WBC:14.89x10^9/L, CRP:76.8 mg/L, Sedimentation:70 mm/hour) and anemia (Hb:10 g/dL, MCV:66.5 fL, RDW:16.3%) were observed

RESULTS

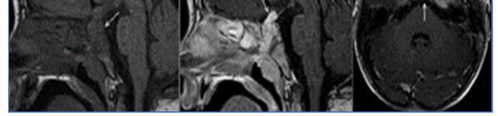
- Water deprivation test was performed with a prediagnosis of diabetes insipidus in a patient with a urine density of 1001
- After the test, the patient was diagnosed with diabetes insipidus with serum osmolarity 303 mOsm/L and urine osmolarity as 121 mOsm/L. Urinary osmolarity increased by 330% after administration of 10 microgram of desmopressin acetate nasal spray solution
- With these findings, the patient was diagnosed with CDI and desmopressin treatment was initiated

Water Deprivation Test

Hour (h)	Body Weight (kg)	Loss of Body Weight (%)	Blood Pressure (mmHg)	Heart Rate (/min)	Serum Sodium (mmol/L)	Serum Osmolarity (mOsm/L)	Urine Osmolarity (mOsm/L)	Urine Density	Urine Volume (mL/h)	Plasma ADH (pmol/L)
1st	17		116/56	116	**139**	**284,6**	**44,7**	**1001**	120	**3,69**
2nd	16,5	%3	104/66	110	141	288,4	54,4	1001	135	3,96
3rd	16,3	%4,2	116/70	117	144	295	59,2	1001	110	
4th	16,2	%4,7	103/75	125	**148**	**303**	**121**	**1004**	**40**	**4,78**
	10 mcg desmopressin was administered intranasally									
5th	16,1	%5,3	105/72	117	145	296,4	245,3	1010	30	
6th	16,1	%5,3	108/72	127	**145**	**296,7**	**407,7**	**1016**	**10**	



Skeletal survey of the patient shows a lytic expansile bone lesion in the mid diaphyseal part of the left clavicle *(white arrows).*



Sagittal plane T1-weighted MR image (a) reveals the absence of posterior pituitary bright spot *(white arrow)*. Note the destruction of sella turcica and the body of the sphenoid bone. Sagittal plane post-contrast T1-weighted MRI (b) demonstrates the thickened enhancing pituitary stalk *(white arrow)*. Also, note the heterogeneous enhancement of the sphenoid bone around the pituitary gland. Axial plane post-contrast T1-weighted MRI of the brain (c) shows the destruction and infiltration of the skull base, more prominently on the sphenoid bone which is enhancing heterogeneously *(white arrows)*.

- A lytic expansile bone lesion in the mid-diaphyseal part of the left-clavicle was found on skeletal survey
- On pituitary imaging, it was observed that the height of the pituitary anterior gland was slightly increased
- The infundibulum was significantly thick and the bright signal of the neurohypophysis was not observed
- On cranial MRI, widespread lytic-destructive bone lesions were observed in the bone structure forming the sella turcica, and the lateral wall of the left orbital
- The patient, who had a pre-diagnosis of LCH, was directed to an external center for bone biopsy and it was learned that the treatment for the diagnosis of LCH was planned

CONCLUSIONS

Presence of isolated CDI with low-grade intermittent fever should be a warning for the diagnosis of LHH. But the patients with CDI should be evaluated in terms of LHH, the most known underlying cause, regardless of the presence of fever.

REFERENCES

1. **Guyot-Goubin A et al.** Descriptive epidemiology of childhood Langerhans cell histiocytosis in France, 2000-2004. *Pediatr Blood Cancer 2008; 51: 71-75.*

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