A case of central diabetes insipidus developed 4 years after the non-CNS-risk unifocal bone lesion of Langerhans cell histiocytosis

Hisae Nakatani1, Kentaro Miyazaki2, Kei Takasawa3, Akira Morimoto4, Masayuki Nagasawa5, Akihiro Oshiba1, Kenichi Kashimada6

1 Department of Pediatrics, Musashino Red Cross Hospital, Tokyo, Japan
2 Department of Pediatrics, Tokyo-Kita Medical Center, Tokyo, Japan
3 Department of Pediatrics and Developmental Biology, Tokyo Medical and Dental University (TMDU), Tokyo, Japan
4 Department of Pediatrics, Jichi Medical University of Medicine, Tochigi, Japan

Background

Langerhans cell histiocytosis (LCH)
- A rare disease with an incidence of less than 10 per million
- Characterized by the clonal proliferation of pathogenic Langerhans cells
- The clinical courses: diverse, ranging from spontaneously remitting single organ disease to life-threatening multisystem involvement
- Complications: diabetes insipidus (DI)

Case presentation

A 6-year-old Japanese boy (Figure 2)
- 2 yrs: A single lytic lesion in his femur → histologically diagnosed as LCH → Clinically self-limited
- 6 yrs: Hospitalized due to polyuria and polydipsia for one and a half months
  - Revealed hyperosmotic dehydration (s-Osm 298 mmol/kg; Table 1) with inappropriately diluted urine (u-Osm 205 mmol/kg) and polyuria (7570 ml/m²/day).
  - Pitressin test: compatible with a diagnosis of CDI. (Table 2)
  - Anterior pituitary functions was intact by stimulating tests.
  - Administration of DDAVP dramatically improved polyuria and polydipsia.
  - Brain MRI (Figure 3): consistent with the diagnosis of CDI due to LCH
  - Examination of spinal fluid; β-hCG not detected

Discussion

The risk for CDI in LCH

- The incidence of DI in overall LCH patients is 12-25%. 1-4
- LCH with CNS-risk lesions (% craniofacial lesions):
  - The risk for DI: High (50%) 4
- Systematically following up by endocrinologists is recommended.
- LCH with non-CNS-risk single-system single site lesion:
  - The risk for DI: Estimated to be extremely low 21
  - A large cohort study of LCH with single-system single site type in Japan (n=146)
  - Association with DI: none 21
- Not routinely followed up by endocrinologists.
- Our case suggests that
  - Even in a patient with non-CNS single organ affected, DI could be involved as a complication of LCH.
  - Further epidemiological studies with an accumulation of cases are necessary.

Conclusion

We recommend to systematically follow up the patients with a history of LCH, even non CNS-risk single-system single site affected type

References