

Central Hypothyroidism: The Endocrine “Memory” for Dyke–Davidoff–Masson Syndrome

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Abstract: Hypothyroidism is a worldwide medical problem, most of the cases being related to primary causes like thyroid autoimmune background or multinodular goiter. A particular form of thyroid hormone insufficiency of the central type is related to Dyke–Davidoff–Masson (DDM) syndrome, which is a severe condition associating hemicerebral hypoplasia or even atrophy, a consequence of a brain injury during the fetal period of time or the first years of childhood. In addition to neurological damage, pituitary function may be affected, requiring hormone replacement.

Keywords: hypothyroidism; Dyke–Davidoff–Masson syndrome; pituitary gland; dwarfism; brain

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Background

Hypothyroidism is a worldwide medical problem, most of the cases being related to primary causes [with or without] thyroid autoimmune background or multinodular goiter [1,2]. A particular form of thyroid hormone insufficiency of the central type is related to Dyke–Davidoff–Masson (DDM) syndrome, which is a severe condition associating hemicerebral hypoplasia or even atrophy, a consequence of a brain injury during the fetal period of time or the first years of childhood [3]. In addition to neurological damage, pituitary function may be affected, requiring hormone replacement [3–5].

Aim

Our purpose is to introduce a case of DDM—related endocrine picture.

Material and Methods

This is a case presentation following medical records based on an endocrine and imaging data.

Results—Case Presentation

This is a 19-year-old male who is admitted for a central hypothyroidism check-up. His medical family history is irrelevant. His personal medical history includes: birth weight of 3500 g (no maternal anomaly was registered) but with neonatal injuries and then a severe meningoencephalitis episode complicated with sepsis and a 10-day coma. Consecutively, he developed neurological anomalies: right hemiparesis, epilepsy (requiring lifelong therapy), and right eye amblyopia. He was confirmed with pituitary dwarfism based on extremely low IGF1 of 9 ng/mL (normal: 136–461 ng/mL) and therapy with growth hormone was offered to the patient but it was not tolerated. By the age of 11, the subject was confirmed with central hypothyroidism and replacement therapy was started. At age of 17 yrs he had a height of 126.8 cm in association with G3P3 pubertal development with intact normal testosterone/FSH/LH assays. On current admission, adequate substitution for thyroid function is found based on TSH of 0.7 μ UI/mL (normal: 0.5–4.5 μ UI/mL), FT4 of 11.85 pmol/L (normal: 9–19 pmol/L) under daily 50 μ g levothyroxine, as well as normal glucocorticoid axes: ACTH of 13 pg/mL (normal: 3–66 pg/mL), plasma morning cortisol of 10.89 μ g/dL (normal: 4.82–19.5 μ g/dL) as well as gonadal axes based on plasma total testosterone of 3.79 ng/mL (normal: 2.49–8.36 ng/mL), FSH of 7 mUI/mL but low IGF1 of 22 ng/mL (normal: 133–395 ng/mL), and persistent since first endocrine admission prolactin deficiency of 0.3 ng/mL (normal: 4–15 ng/mL). Magnetic resonance imaging and computed tomography confirmed left cerebral hemiatrophy with liquid transformation (very suggestive for DDM syndrome), corpus callosum agenesis, and empty sella. Further levothyroxine substitution is needed in addition to neurological medication.

Conclusions

DDM-related pituitary insufficiency selectively affecting thyroid axes requires lifelong replacement.

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Conflicts of Interest: The authors declare no conflict of interest.

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