

An unusual presentation of Hashimoto thyroiditis (HT) and precocious puberty: The VAN WYK-GRUMBACH Syndrome

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INTRODUCTION:

The association of primary hypothyroidism and isosexual precocious pseudopuberty in females was first described in 1960 by Van Wyk and Grumbach. The unique elements that lead to the diagnosis are:

- Hashimoto's hypothyroidism
- FSH-dominated sexual precocity
- Non advanced bone age

AIM:

Describe an 8.5yrs old girl with hypothyroidism due to HT and clinical and hormonal features of Van-Wyk and Grumbach syndrome.

CASE REPORT:

An 8.5 yrs girl was referred to the endocrinology department with a suspicion of precocious puberty. She had no history of visual disturbance or headaches but was overweight for her height. On examination:

- Height: 130.6cm (50th centile), MPTH: 159.5 (25thcentile)
- Weight: 30Kg (75th centile)
- Tanner's stage B 2-3, PH 1, AH 1
- Hormonal investigations:
 - TSH: 32.46mIU/ml (0.4-5)
 - FT4: 0.98/ng/dl (0.9 – 1.9)
 - Abs anti TPO: 379.2IU/ml(<16) and anti TG: 543.6IU/ml (<100)
 - LHRH testing showed an FSH-dominated, prepubertal response with a 0-60 min FSH rise from 2.17 to 6.01 mIU/ml and an LH rise from 0.38 to 2.54mIU/ml. Oestradiol: 40.7pg/ml (NR <27).
 - Additional hormonal tests: PRL: 21.8 ng/ml (4.8 – 23.3), Testosterone: 2ng/dl (2-20), DHEA-S: 69mcgr/dl (2.8 -85.2), 17-OHPRG: 0.57ng/ml (0.2-0.5), β -HCG: <1U/lt (<5) and α -FP levels:<2ng/ml (<15ng/ml).
 - Pelvic ultrasound revealed a pubertal uterus (37x10x20mm) and multicystic ovaries (RO: 5.4cc, LO:4.5cc) with multiple dominant follicles.
 - Bone age was equal to her chronological age, in contrast to the pubertal Tanner stage 2-3 and the ultrasound
 - TTT: Thyroxine increased gradually to 50mcg/day (1.7mcgr/Kg). Clinical, hormonal and ultrasound findings returned to normal without additional therapy.

CONCLUSIONS:

Hypothyroidism (either isolated or in the context of autoimmune thyroiditis), although usually predisposes to delayed puberty it may also lead to isosexual precocious pseudopuberty. Recognition of this syndrome is important because thyroid hormone substitution completely resolves symptoms and hormonal abnormalities.

Bibliography

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