



EXAGGERATED ADRENARCHE AND EXOGENOUS OBESITY: A DIAGNOSTIC CHALLENGE

Ferreira M ; Semer B; Queiroz E; Brigatti N; Athayde D; Pinheiro, C; Steinmetz L; Cominato L; Menezes Filho H; Kuperman H; Setian N; Damiani D Instituto da Criança - Hospital das Clínicas - Faculdade de Medicina - Universidade de São Paulo

The autors certify that there is not any conflict of interests in this manuscript.

INTRODUCTION

Exaggerated adrenarche is an extreme variant of the maturation of the reticular zone of the adrenal cortex, often associated with hyperinsulinemia and obesity. Hyperandrogenism by congenital adrenal hyperplasia and adrenal neoplasms are differential diagnoses.

CASE REPORT

Male, 8 years and 3 months, came from another service with diagnosis of precocious puberty and obesity, already being treated with Leuprolide acetate for 1.5 years. His complaint was weight gain and growth of body hair since 5 years old. Physical examination: cushingoid facies, weight 52.5 Kg (BMI 35.9 – Z = +4.87), height 121.5 cm (Z = -1.24), abdominal circumference 99cm, acne, gynecomastia, acanthosis nigricans, Tanner stage P5G2, testicles 3 cm³.

Initial exams: Bone age of 10 years (75% mature height), ACTH 43 pg/mL, cortisol 7.6 mcg/dL, urinary cortisol 576mcg/24h (92-478), plasma renin activity 1.2ng/ml/h (0.2-2.8), DHEAS 4080ng/mL (244-2470), adrenal ultrasound, abdomen CT and pituitary gland MRI were normal. Lab work-up:, ACTH test results: DHEAS T0 = 4080ng/mL, 4260ng/mL, Androstenedione T0 = 3.2 ng/mL T60 = 4.6 ng/mL, T60 =170HProgesterone T0 = 0.6 ng/mL T60 = 7.2 ng/mL (0.59 - 3.44).

It was suggested the hypothesis of Nonclassical Congenital Adrenal Hyperplasia (CAH) due to 3ß-Hydroxysteroid Dehydrogenase and therapy with corticosteroids was started, but turned out unsuccessful. Meanwhile, results of ACTH - estimulated 170HPregnenolone = 1719 ng/dL (<3000), cortisol (F) = 14 mcg/dL, with 170H Pregnenolone/F ratio = 0.12 (<67) ruled out this diagnosis. (3,4) Then, exaggerated adrenarche associated with exogenous obesity became the main hypothesis and patient started treatment with metformin for insulin resistance (HOMA-IR = 7.1), aromatase inhibitor due to the advanced bone age (BA = 13y6m CA = 10y7m) and GH replacement.

The patient reached the final height of 161 cm (Z= - 2.03), in accordance with the prediction, yet below his target height (173.7; Z = -0.33).





CONCLUSION

Exaggerated adrenarche is considered a diagnosis of exclusion and its association with obesity is becoming more common in the pediatric and adolescent population, which brings a concern for the medical community in terrms of treatment options. It may be difficult to make a decision between no medication ("adrenarche") and aromatase inhibitors plus rhGH. Our patient had a final height below his TH, as opposed to a normal adrenarche that usually do not compromise final height. Further studies are necessary to better clarify this condition.

REFERENCES:

- 1. W.F. Paterson et al. Exaggerated adrenarche in a cohort of Scottish children: clinical features and biochemistry. Clinical Endocrinology (2010) 72, 496–501. doi: 10.1111/j.1365-2265.2009.03739.x
- 2. Pang, S. Congenital adrenal hyperplasia owing to 3 beta-hydroxysteroid dehydrogenase deficiency.. Endocrinol Metab Clin North Am. 2001 Mar;30(1):81-99, vi-vii.
- 3. Lutfallah, C et al. Newly Proposed Hormonal Criteria via Genotypic Proof for Type II 3β-Hydroxysteroid Dehydrogenase Deficiency. The Journal of Clinical Endocrinology & Metabolism 2002 87:6,2611-2622

DOI: 10.3252/pso.eu.55ESPE.2016

4. Sperling, M. Adrenal Cortex and Its Disorders. In: Sperling, M. (Ed.). *Pediatric Endocrinology* (pp. 471-526). 4.ed. Philadelphia, PA: Elsevier, 2014.









