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Case Report

The Case of a Patient with Primary Amenorrhea and Arterial Hypertension

Alvaro Pachón-Burgos[1], Don Manuel Guerrero-Monteza[2], Rafael Santamaria-Alvarez[3], Johan Serrano-Quintero[4] and Martha Patricia Chan-Aguilar[5]*

[1]*Department of Internal Medicine and Endocrinology, Hospital Regional de David Rafael Hernández Loeche, Panamá*

[2]*Department of Urology, Hospital Regional de David Rafael Hernández Loeche, Panamá*

[3]*Department of Pathology, Hospital Regional de David Rafael Hernández Loeche, Panamá*

[4]*Department of Internal Medicine, Hospital Regional de David Rafael Hernández Loeche, Panamá*

[5]*Department of Pediatrics and Neonatology, Hospital Jose Domingo De Obaldía, Panamá*

Abstract

Objective: To highlight the importance in the etiological study of secondary arterial hypertension.

Method: Describe a clinical case: clinical history, physical examination, laboratory and radiographic studies.

Results: A 17-year-old girl came to our endocrinology unit with primary amenorrhea, absence of secondary sexual characteristics, resistant hypertension and upper left abdominal discomfort. Her sister died by a similar health problem. Her blood pressure is 240/140 mmHg, and she needs five drugs for hypertension control, her height is 1.74 m and her weight is 54 kilograms, she has eunuchoidism, lack of breast development, lack of axillary and pubic hair and blind vagina. Her laboratories show us: hypokalemia 3.3 mEq/L, metabolic alkalosis, almost undetectable levels of estradiol and testosterone with very high levels of folliclestimulating hormone (94 milli-international units/ml) and luteinizing hormone (74.8 milli-internationalunits/ml), a hypergonadotropic hypogonadism, low levels of 17OH progesterone 0.17 ng/ml (0.1-1.2),11deoxycortisol 127 ng/dL (<127), cortisol 2 ug/dL (5-25), 17-OH pregnenolone 405 ng/dL (950),DHEA-SO4 15 ug/dL (35-430),aldosterone 2 mg/dL (0-28), renin plasma activity 0.14 ng/mL/h (0.25-5.82) and very high levels of pregnenolone 225 ng/mL (0.46-1), corticosterone 11090 ng/dL (59-1293), deoxy-corticosterone 494ng/dL (<18)and ACTH 365 pg/mL (0-46). Genotype analysis: XY. Computed axial tomography (CAT) of abdomen and pelvis showed enlarged adrenal glands with the presence of a large adenoma in left adrenal gland that displaces the left kidney down.

Conclusions: All clinical data suggest congenital adrenal hyperplasiadue to 17 alpha-hydroxylase deficiency.

Keywords: Amenorrhea; Congenital adrenal hyperplasia; Hypertension

Case Presentation and Conclusion

A 17-year-old girl come to our endocrinology unit with primary amenorrhea, absence of secondary sexual characteristics, resistant hypertension (she needs five drugs for hypertension control) and upper left abdominal discomfort. Her blood pressure is 240/140 mmHg, her height is 1.74 m and her weight is 54 kilograms, she has eunuchoidism, lack of breast development, lack of axillary and pubic hair and blind vagina. Her sister died by a similar health problem. Her laboratories showed us: hypokalemia 3.3 mEq/L, metabolic alkalosis, almost undetectable levels of estradiol and testosterone with very high levels of follicle stimulating hormone (94 milli-international units/ml) and luteinizing hormone (74.8 milli-international units/ml), a hypergonadotropic hypogonadism, low levels of 17OH progesterone 0.17 ng/ml (0.1-1.2), 11deoxycortisol 127 ng/dL (<127), cortisol 2ug/dL (5-25), 17-OH pregnenolone 405 ng/dL (950), DHEA-SO4 15 ug/dL (35-430), aldosterone 2 mg/dL (0-28), renin plasma activity 0.14 ng/mL/h (0.25-5.82) and very high levels of pregnenolone 225 ng/mL (0.46-1), corticosterone 11090 ng/dL (59-1293), deoxy-corticosterone 494 ng/dL (<18) and ACTH 365 pg/mL (0-46). Genotype analysis: XY. An abdominal and pelvic ultrasound found a large mass in the anatomical region of the left adrenal gland, absence of uterus, fallopian tubes and ovaries. Computed Axial Tomography (CAT) of abdomen and pelvis showed enlarged adrenal glands with the presence of a large adenoma in left adrenal gland that displaces the left kidney down Figure 1. We did surgical resection of intraabdominal testes and left adrenalectomy and we found a giant myelolipoma of 10x6x4 cm and abdominal atrophic testes in the histopathological study Figure 2. All clinical data (medical history, physical examination and laboratory results) suggest congenital adrenal hyperplasia [1] due to17 alpha hydroxylase deficiency. After one year of follow-up she does not have hypertension and she just needs dexamethasone 0.25 mg per day. Sporadic myelolipomatous lesions in the adrenal hyperplastic tissue have been noted before in two cases [2,3]. Interestingly, a large adrenal myelolipoma in a case of 17 alpha-hydroxylase deficiency has been described, which suggests pathological association of this tumor with prolonged ACTH stimulation.

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***Corresponding author:** Alvaro Pachón-Burgos, Department of Internal Medicine and Endocrinology, Hospital Regional de David Rafael Hernández Loeche, Chiriquí, Panamá, Tel: (507)66152440; E-mail: alvarop5@yahoo.com

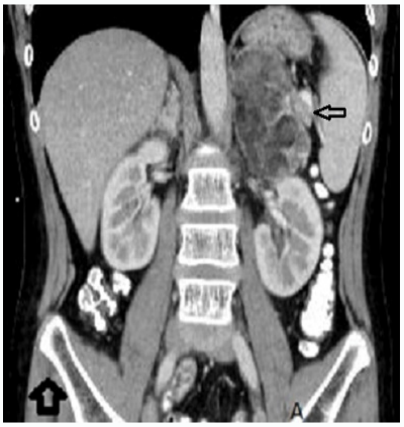

Figure 1: Computed axial tomography (CAT) of abdomen and pelvis showed enlarged adrenal glands with the presence of a large adenoma in left adrenal gland that displaces the left kidney down.

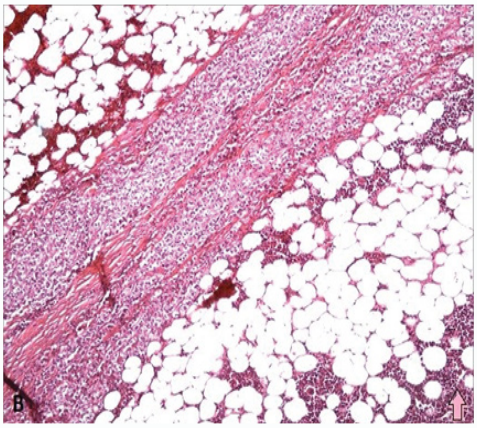

Figure 2: Surgical resection of intraabdominal testes and left adrenalectomy and we found a giant myelolipoma of 10 cm × 6 cm × 4 cm and abdominal atrophic testes in the histopathological study.

References

1. Miller WL, Auchus RJ. The Molecular Biology, Biochemistry, and Physiologyof Human Steroidogenesis and Its Disorders. Endocr Rev. 2011;32(1):81-151.
2. Yanase TO, Simpson ER, Waterman MR. 17a-Hydroxylase/17,20-Lyase Deficiency: From ClinicalInvestigation to Molecular Definition. Endocr Rev. 1991;12(1):91-108.
3. Merchant SH, Herman CM, Amin MB. Myelolipoma Associated With Adrenal Ganglioneuroma. Arch Pathol Lab Med. 2002;126(6):736-7.