Urology Case Reports 18 (2018) 57–59

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Andrology and fertility

Case report: Clitoromegaly as a consequence of Congenital Adrenal Hyperplasia. An accurate medical and surgical approach

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ARTICLE INFO

Article history:
Received 23 January 2018
Accepted 14 February 2018
Available online 13 March 2018

Keywords:
Congenital Adrenal Hyperplasia (CAH)
Clitoromegaly
Surgical techniques

ABSTRACT

We present a case of a woman with a history of Congenital Adrenal Hyperplasia (CAH) diagnosed at the age of 12, who was referred to our unit for surgical treatment. Despite the initial diagnosis was an indirect inguinal hernia, it was a misdiagnosis. Once in our service, this was corrected into clitoromegaly secondary to CAH. Physical examination and imaging test discarded other abnormalities, such as secondary effects androgenization. Regarding surgical treatment, the techniques used were Spencer and Allen combined with Kumar, which are the most used for clitoroplasty but also less used in Peru.

Introduction

Congenital Adrenal Hyperplasia (CAH) is an increase in size and metabolic activity of the adrenal cortex or medulla due to inherited enzymatic disorders.[1] The enzymes implicated in this disorder are steroid 21-hydroxylase (21-OH), 11β-Hydroxylase and 3β-hydroxysteroid dehydrogenase.[2] This rare condition is present in 1 of 15,000–16,000 live births worldwide[1,2] and it's caused by an increase in biosynthesis of androgens such as testosterone and 5α-dihydrotestosterone which are the ones to account for the clinical presentation of this disease which are virializing genitalia in women.[1] The main management is either surgical or medical treatment with the normalization of the genitalia as the objective.[1,2]

Case presentation

A 18 – years - old female was referred to Gynecology service to discard a suspected indirect inguinal hernia. She was diagnosed at the age of 12 and has been treated with Dexamethasone 0.5 mg per night every day since her diagnosis. Physical examination was under normal conditions, including genitourinary exam, except for an increased size of clitoris, approximately 3 cm (Fig. 1). Regarding the imaging tests, the pelvic ultrasound showed a normal uterus. Likewise, both laboratory test and karyotype were normal, with the only exception of high levels of Androstenedione. Finally, her surgical risk was done, and both ASA and Goldman scale were at stage I.

The patient was admitted to the Gynecologic inpatient service for her clitoroplasty. The chosen procedure was a Spencer and Allen, it started with the circumcision from 9 to 3 following a clock pattern, leaving 5 mm of skin. Then, the cavernous and clitoris planes were dissected, then, the neurovascular bundle was isolated, followed by the cavernous body isolation around 4 cm approximately (Fig. 2). All of this was done aimed at cutting 3 cm of the cavernous body so as preserve part of the base of it and its to neurovascular bundle. Next, the distal and proximal planes were sutured with Vycril 2.0, then the surgeons fixed the Buck fascia to the periosteum-pubic fascia using the clock pattern of 9, 11, 1, and 3. Lastly, the closing of skin and proper hemostasia were done (Fig. 3).

Discussion

People with CAH usually presents as an asymptomatic condition; although, only 11% of all the patients develops clitoromegaly.[1] The first step in the diagnosis is the dosage of precursor hormones, such as 17-hydroxyprogesterone (17-OHP), cortisol and aldosterone to account the deficiency of the enzymes.[2] With this tests results, the physician also verifies if the patient has cortisol insufficiency, to classify the patient as a classical or a non-classical presentation of this disorder,[2] being the non-classical form the most common of the two.[1] In this particular case, the diagnosis was made by dosage of the hormone final products such as DHEA-s and

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https://doi.org/10.1016/j.eucr.2018.02.011

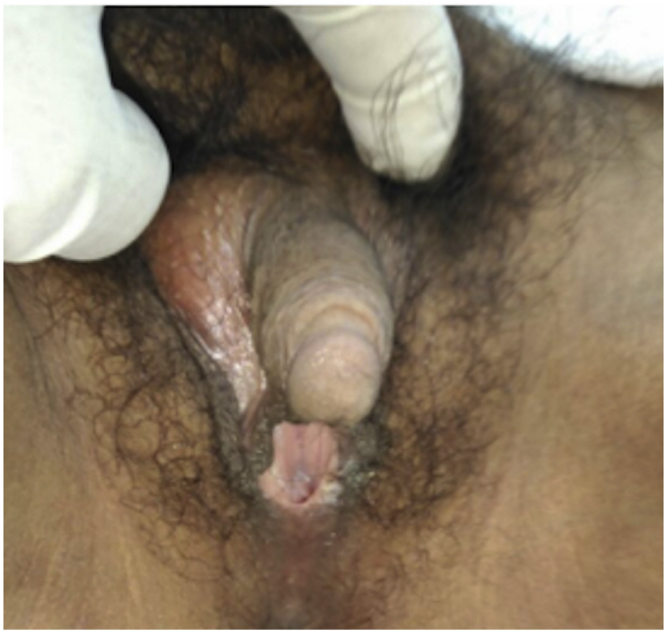

Fig. 1. Clitoromegaly before the surgery.

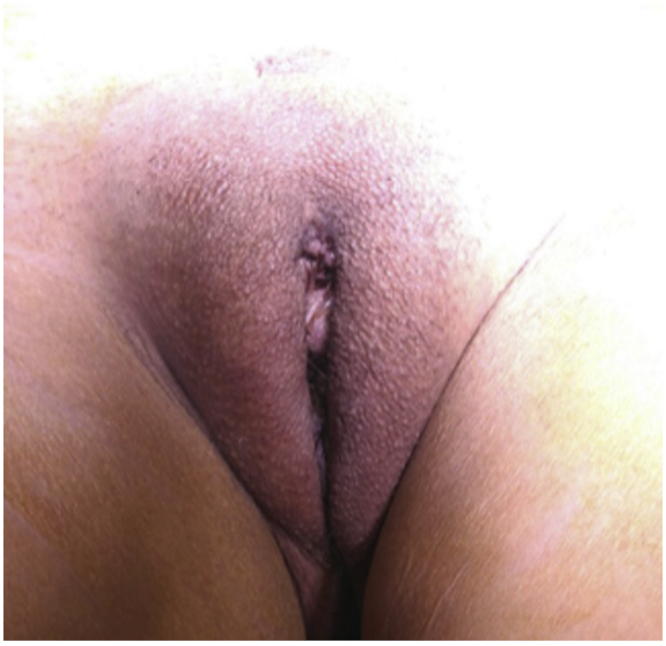

Fig. 3. Genitalia after the surgical procedure.

Androstenedione but tests for 21-OH or 17-OHP were not made. Although this are used for the diagnosis, this are not usually taken in Peru.

There are many techniques which have been used for people with disorders of sexual development secondary to a Congenital Adrenal Hyperplasia condition. One of them is the enlargement of clitoris. The surgical approaching of this is based on many techniques aimed to create normal looking female external genitalia,[3] and focused on fixing the volume and size of clitoris such as Young, Lattimer, Barinka, Fonkalsrud, Lanzuela, Spencer & Allen, Kumar, Kogan, etc.[4] One the most successful is Spencer & Allen, and Kumar techniques which are focused on preserve the clitoris sensitivity.[4] The surgeons must decide the proper technique and what does the patient want to.

In this case, the surgical procedure was a combined procedure between the Kumar and the Spencer and Allen techniques, these are one of the most complete and esthetic procedures and consist in preserve the base of clitoris joined to the neurovascular bundle and the Buck fascia instead of skin, preserving the sensitivity and giving an esthetic profile.[4] Many outcomes for clitoroplasty reported good results following this procedure, such as Milan cases, which showed 100% success in terms of reduction and cosmesis.[5]

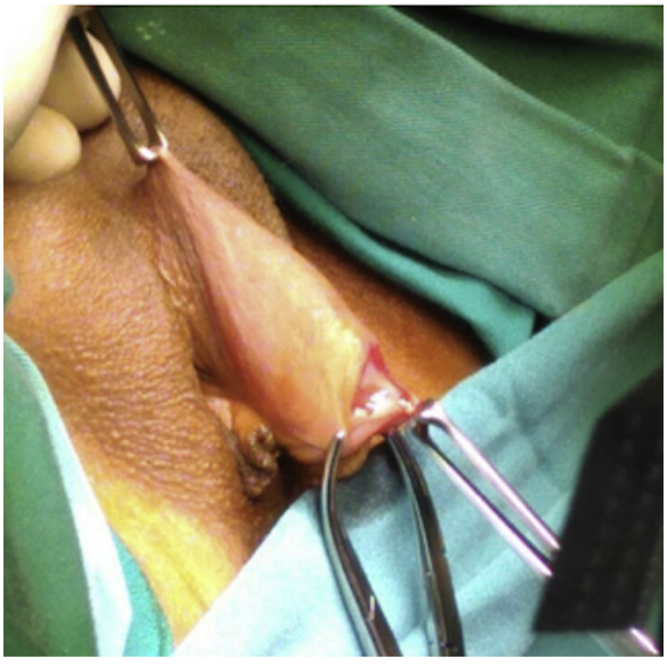

Fig. 2. Clitoroplasty.

Conclusions

To conclude, our case demonstrated important clinical and surgical lessons. First, it is important to highlight the correct approach of CAH and its workup using the correct tests for rule out diseases secondary to this condition. Likewise, it is also important to consider the correct surgical approach, insomuch as imperative to take into account how comfortable will the patient feel after surgery.

Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.eucr.2018.02.011.

References

1. Carmina E, Dewailly D, Escobar-Morreale H, Kelestimur F, et al. Non-classic congenital adrenal hyperplasia due to 21-hydroxylase deficiency revisited: an update with a special focus on adolescent and adult women. *Hum Reprod Update*. 2017;23(5):580–599.
2. Turcu A, Auchus R. Adrenal steroidogenesis and congenital adrenal hyperplasia. *Endocrinol Metab Clin N Am*. 2015;44(2):275–296.
3. Yanovik F, Cherian A, Steven L, Mathur A, Cuckow P. Current practice in feminizing surgery for congenital adrenal hyperplasia; A specialist survey. *J Pediatr Urol*. 2013;9:1103–1107.

4. Nuñez y col. Reconstruccion genital intedral en la hyperplasia suprarenal Congenita: sensibilidad, estetica y funcion (embarazo). *Cir Plast Iberlatinoam*. 2010;36(1):79–86.

5. Van der Toorn F, et al. Introducing the HOPE (Hyypospadias Objective Penile Evaluation)- score: a validation study of an objetive scoring system for evaluating cosmetic appearance in hypospadias patients. *J Pediatr Urol*. 2013;10, 1—1.