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Clitoromegaly due to an epidermal inclusion cyst: A case report

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ABSTRACT

Background: Clitoromegaly is often a sign of androgen excess; however, non-hormonal causes must be ruled out. We report the case of an adolescent with isolated clitoromegaly without clinical or biochemical evidence of hyperandrogenism.

Case: A 16-year-old female was referred due to a clitoromegaly of 12 months of evolution. Examination of the pubic region revealed normal female genitalia with an enlarged clitoris, 4 cm long and 2.5 cm wide. The clitoris was painless, soft on palpation, and mobile over deeper layers. There were no signs of virilization, and the patient did not report dysuria or difficulties with sexual intercourse. Her medical record was also unremarkable, with no female circumcision, family history of birth defects, or genital abnormalities. Hormone profile blood tests were normal. Pelvic ultrasound examination was normal, but a high-resolution scan with a linear transducer confirmed the presence of a cyst, lying anterior to the clitoral body and glans. The cyst was surgically removed with special care to preserve the clitoral neurovasculature. The pathological report disclosed an epidermoid clitoral cyst. The patient described emotional well-being, satisfactory sexual function, and no discomfort after a year of follow-up.

Conclusion: Epidermal clitoral cysts represent an unusual cause of clitoromegaly. These cysts should be ruled out as a differential diagnosis after an exhaustive semiological and endocrinological examination.

1. Introduction

Clitoromegaly is often a sign of virilization caused by androgen excess [1]. Although rare, non-hormonal causes such as benign neoplasms require differential diagnosis [2]. We report the case of an adolescent with isolated clitoromegaly without clinical or biochemical evidence of hyperandrogenism.

2. Case Presentation

A 16-year-old female consulted the department of endocrinology with a referral for evaluation of clitoromegaly. The enlargement of the clitoris was noticed about a year previously as a painless mass that did not cause dysuria or difficulties with sexual intercourse. Upon referral she reported emotional discomfort over having to wear loose clothing

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due to shame of the condition being evident.

She had hidden this problem from her parents until she had a bicycle accident. At the hospital, physicians disclosed her problem during the physical examination. There was no evidence of female circumcision, a family history of inborn defects, or genital abnormalities, and her medical record was also unremarkable. She denied history of any medication or androgen use. Her menarche occurred when she was 11, with menstrual cycles every 35 days.

Upon physical examination, she had well-developed female sexual characteristics, and normal body mass index and blood pressure. Thyroid palpation was normal and there were no signs of virilization (i.e hirsutism, deep voice, or male-pattern alopecia). We found no *café-au-lait* spots or freckles in the armpits. Her breasts were Tanner stage 4, and there was no galactorrhea. No abdominal masses were palpable. Inspection of the vulvar region revealed an enlarged clitoris, 4 cm long and 2.5 cm wide (Fig. 1). The clitoris was soft on palpation and mobile over deeper layers with well-formed glans. The urethral and vaginal orifices were normal. The labia majora and minora were normal in appearance, and pubic hair was Tanner stage 4. Speculum examination revealed a well-estrogenized vaginal mucosa and a normal-appearing cervix; bimanual examination was unremarkable.

The patient's hormone profile was normal, including luteinizing hormone, follicle-stimulating hormone, estradiol, total testosterone, androstenedione, dehydroepiandrosterone sulfate and 17-hydroxyprogesterone, nocturnal salival cortisol, thyroid function tests and prolactin. Gynecological ultrasound examination was normal, but the high-resolution scan with a linear transducer confirmed the presence of a cyst in the clitoral region with homogeneous and dense content (Fig. 2).

Surgical removal was performed under regional anesthesia with special care to preserve the clitoral neurovasculature. There were no complications during the postoperative period and the pathological report revealed an epidermoid clitoral cyst (Fig. 3). After one year of follow-up, sexual function was satisfactory, discomfort resolved, and the patient reported emotional well-being.

3. Discussion

Clitoromegaly is the enlargement of the clitoris [1], with most cases seen in adult women. Reports among adolescents are rare. Brodie et al. [2] published data from 58 patients of different pediatric ages in order to create standard references for the anterior vulval structures, and examined the relationship between the clitoral hood and labia minora. They reported a mean clitoral diameter of 4.5 mm and mean length of 20.9 mm in girls aged 13–16 [2].

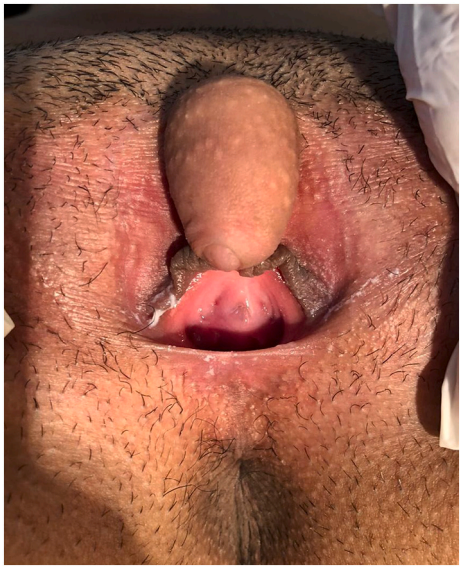

Fig. 1. Large mass of the clitoral hood resembling a phallus and hiding the clitoris.

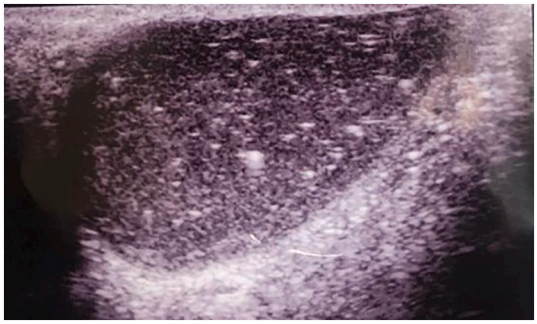

Fig. 2. High-resolution ultrasonography shows a circumscribed oval-shaped mass, with a thin wall, echogenic reflections and no Doppler flow, consistent with cystic image measuring 34 × 12 × 20 mm.

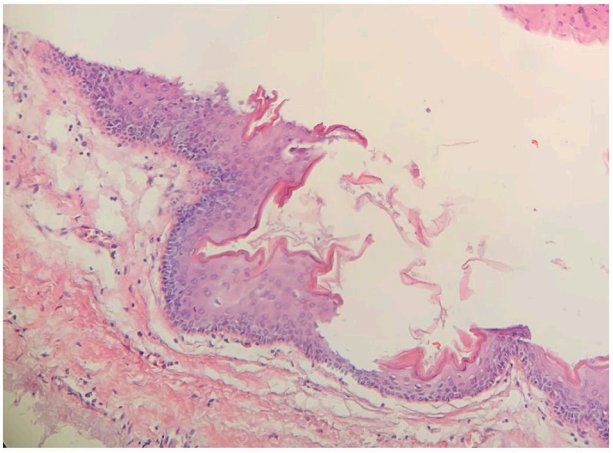

Fig. 3. Histopathology determined an epidermal cyst lined by stratified squamous epithelium with a granular layer and filled with keratin flakes.

Clitoromegaly can be congenital, frequently diagnosed at birth, or acquired, which can have hormonal, and non-hormonal causes. Hormonal-induced clitoromegaly may be caused by hyperandrogenism, polycystic ovary syndrome, virilizing ovary or adrenal tumors, and the exposure to exogenous androgens. Non-hormonal causes are produced by neoplasms [1,3].

Androgen excess may occur during intrauterine life. Congenital adrenal hyperplasia is the most common cause of fetal origin. Fetal 21-hydroxylase enzyme deficiency causes glucocorticoid and mineralocorticoid deficiency as a cause of excessive androgen synthesis. The clinical expression will depend on the severity of enzyme alteration, ranging from neonatal hyponatremic dehydration to different degrees of masculinization. The latter ones are due to fetal hyperandrogenism in the classic form. Non-classical forms do not present saline loss or prenatal virilization though they can present clinically with hyperandrogenism during childhood, adolescence, or even adult life. Less common causes are those produced by maternal hyperandrogenism during pregnancy, such as luteoma, hyperreactio luteinalis, virilizing tumors, or consumption of preparations containing androgens. The deficiency of placental aromatase enzyme produces an accumulation of androgens with virilization of the mother and fetuses as an example of androgenization *in utero*, from placental origin [3,4].

Androgen excess in postnatal life, with an abrupt onset, signs of virilization, and high serum androgen titers, is generally related to a tumor of ovarian or adrenal origin. It is mandatory to ask about the use

of exogenous androgens. In the present case, all the above-mentioned causes were ruled out [5,6].

Neurofibromatosis and tumors, among others, must be excluded as non-hormonal causes of clitoromegaly [3]. Neurofibromatosis is an autosomal dominant disorder in which patients present overall characteristic skin lesions (*café-au-lait* spots) and soft-tissue tumors arising from the neural sheath. Isolated clitoral involvement related to this pathology is rare [7,8]. Epidermoid cysts represent a rare cause of non-hormonal clitoromegaly. They are composed of keratinized squamous epithelium invaginated into the dermis or subcutaneous tissue. Cases reported in the international literature have been related to genital mutilation or female circumcision [9–11]. Despite being illegal in most countries, these practices are still being performed among girls from some African and Muslim countries, with epidermal inclusion cysts being a late complication of such intervention. There are few reported cases of epidermal inclusion cysts of the clitoris in which there was no history of trauma or ritual circumcision [12,13,14]; ours therefore contributes to the international literature. Due to the great variety of tumors that can affect the vulva, the histopathological study is essential to confirm the diagnosis.

4. Conclusion

The first step in the management of a patient with clitoromegaly is to rule out endocrinological causes. Secondly, malignant or benign clitoral lesions should be considered. Our case highlights the fact that an epidermoid cyst should be kept in mind as a rare non-hormonal cause of clitoromegaly. Imaging studies could help diagnose unclear situations. Special care of clitoral vascularization and innervation should be taken during the surgical removal of the cyst.

Contributors

Carolina Fux-Otta is the primary author, who performed the endocrinological evaluation and clinical follow-up of the patient; and conducted the literature search and prepared the initial draft of the case report.

Margarita Fuster performed the surgical intervention.

Noelia Ramos performed the endocrinological evaluation and assisted with literature search.

Cristina Trezza performed the histological study and contributed to literature review.

Mónica Ñañez contributed to literature review, language revision and edits of the manuscript.

Ismael Fonseca performed the histological study and contributed to literature review.

Néstor Dicuatro performed the surgical intervention.

Mariana Di Carlo performed the endocrinological evaluation and assisted with literature search.

Carla Bongiorni performed the histological study and contributed to literature review.

José Ochoa performed the imaging diagnosis and literature review.

Otilio Rosato performed the surgical intervention.

Peter Chedraui contributed to literature review, language revision and edits of the manuscript.

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Patient consent

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Conflict of interest statement

The authors declare having no conflict of interest regarding the publication of this case report.

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