CASE REPORT

sciendo

10.2478/AMB-2024-0009

OPERATIVE TREATMENT OF IDIOPATHIC ISOLATED CLITOROMEGALY – A CLINICAL CASE

S. Stoyanov[1], K. Ivanova[2], I. Koleva[1]

[1]Department of Obstetrics and Gynecology, Medical Faculty, Trakia University – Stara Zagora, Bulgaria
[2]Department of General and Clinical Pathology, Medical Faculty, Trakia University – Stara Zagora, Bulgaria

Abstract. *Idiopathic clitoromegaly is a relatively rare disease. A case of isolated idiopathic clitoromegaly is presented. It concerns a 31-year-old woman, in whom the clitoris has showed a tendency for progressive enlargement in the last 3-4 years. The patient had no gynecological and systemic diseases and disorders. Karyotype and hormone levels were normal. No cystic changes in the ovaries and other changes in the abdominal organs were detected during ultrasound. Computerized axial tomographic (CAT) scan of the adrenal glands was performed and showed normal appearance. Method of selection in such cases is operative treatment – clitoroplasty with storage of neurovascular bundle of the clitoris.*

Key words: *clitoromegaly, clitoroplasty*

Corresponding author: *Koni Ivanova, MD, PhD, e-mail: koni_ivanova@yahoo.com*

Received: *14 February 2023,* ***Revised/Accepted:*** *06 February 2024*

INTRODUCTION

Clitoromegaly is a common anomaly, but acquired isolated clitoral elongation is rare [1] and its causes can be: hormonal, non-hormonal, pseudoclitoromegaly, idiopathic clitoromegaly [2, 3].

The degree of clitoromegaly is most often determined according to the classification of Von Prader A., 1975 [2], which, depending on the intensity of hyperandrogenism, differs in several forms: clitoral hypertrophy with reduced vaginal passage; with a narrowed vaginal entrance; marked clitoral hypertrophy, male external genitalia with hypospadias and hypertrophied penis-like clitoris and labia majora like scrotum. Endocrinopathies, virilizing tumors, and androgen use are the most common hormonal causes of clitoromegaly [4].

A case of a patient with isolated idiopathic clitoromegaly treated surgically is presented.

CASE DESCRIPTION

A 31-year-old female, 0-gravida, complained that in the last 3-4 years, the clitoris has shown a tendency for progressive growth. The patient associated the onset of complaints with the beginning of regular sexual intercourse, which became the cause of negative psycho-emotional feelings and physical sensations (a feeling of worry at the thought of the presence of this problem), physical (discomfort with certain movements) and sexual (discomfort and awkwardness during sexual intercourse) discomfort. The woman denied the use of drugs, psychotropic and hormonal medications. No family history was reported – there are no other cases of clitoral enlargement in the family. Appearances of menarche when she was at the age of 15, the menstrual cycle was regular – every 30-32 days, lasts 3-4 days, settled suddenly. From the objective research – the patient was with normal female phenotype. From the gynecological

examination - in a calm state, the clitoris was 20 mm long (according to the patient's data, it increases to 30 mm during sexual excitement) (Figure 1). Vaginal entrance – was with normal anatomy structure. The external opening of the urethra was located 1.5 cm above the entrance to the vagina. The woman has normal secondary female genitalia with female pubic hair. There was no evidence of obesity – height 172 cm, weight 65 kg.

From the analysis of hormonal tests such as: estradiol, progesterone, FSH, LH, DHEA-S, testosterone, Prolactin, ACTH, cortisol, TSH – were in normal referent value, and tumor marker CEA were in normal referent value also. The values in Table 1 showed the hormonal status of the patient.

Table 1. hormonal status of the patient

FSH (follicular phase)	4.18 mIU/ml
LH (follicular phase)	5,03 mIU/ml
Estradiol (follicular phase)	314,6 pmol/l
Progesteron (luteal phase)	25 nmol/l
DHEA-S	9,7 nmol/l
Prolactin	312,3 mU/ml
ACTH	26,3 pg/ml
Cortisol	218 nmol/l
TSH	2,6 mUI/l
testosterone	2,46 nmol/l

Ultrasound of the small pelvis and abdominal organs showed that there was no data for polycystic changes in the ovaries, no tumor formations were visualized in the pelvis.

From the obtained results, no connection could be made between clitoromegaly and the results from the paraclinical and instrumental examinations. The patient underwent clitoroplasty with preservation of the neurovascular bundle (Figure 2 and Figure 3).

OPERATIVE TECHNIQUE

Under general anesthesia, a semilunar incision was made over the dorsal surface of the clitoris. The dorsal surface was prepared to the pubic fascia without cutting the suspensory ligaments. 5 superficial ligatures were applied (the neurovascular bundle was preserved) on the pubic fascia – at 10, 11, 12, 1 and 2 o'clock. They were tied so that there is no sharp angle when tightening them, which makes erection difficult. Excess skin was cut away. With single resorbable sutures, the normal anatomy was restored – the skin was sewn to the foreskin. The patient was followed for a period of 6 months. The postoperative period went smoothly, without early and late complications (Figure 4).

At the follow-up examination 6 months after the intervention, the patient said that the sensitivity of the clitoral area was normal. The woman was satisfied with the aesthetic and functional result (Figure 5).

DISCUSSION

Clitoroplasty is the method of choice in the treatment of clitoromegaly [4,5]. It aims to improve the psychoemotional state of the woman. At the same time, it is necessary to preserve sexual function and sensitivity with a mandatory cosmetic effect.

For the first time, Young performed a similar plastic correction on a child with congenital adrenal hyperplasia in 1937 [6]. Depending on the intensity of

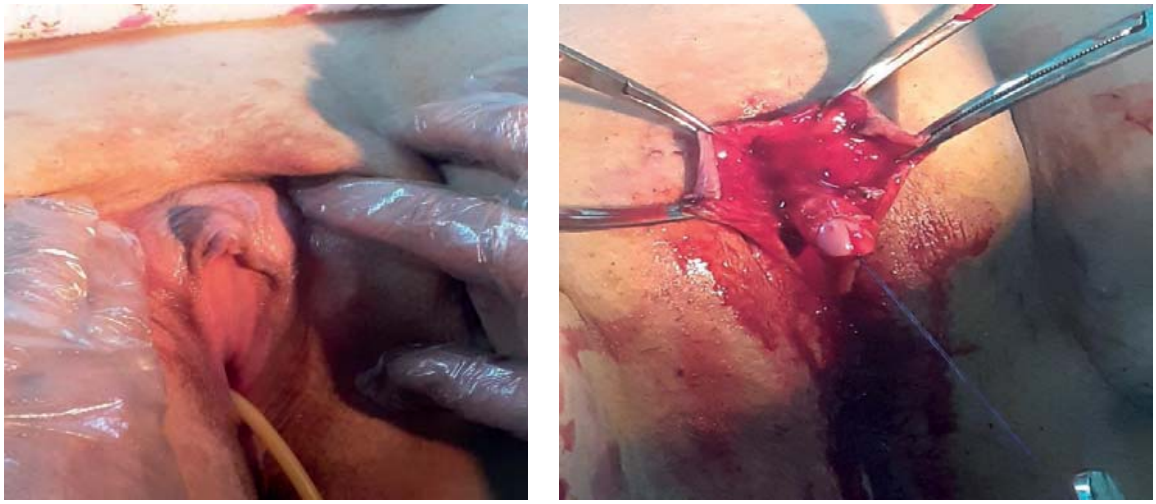

Fig. 1, 2. Clitoris, labia minora, urethral entrance and vaginal entrance before surgical correction (left); Dissection of the foreskin from the dorsal surface of the clitoris (right)

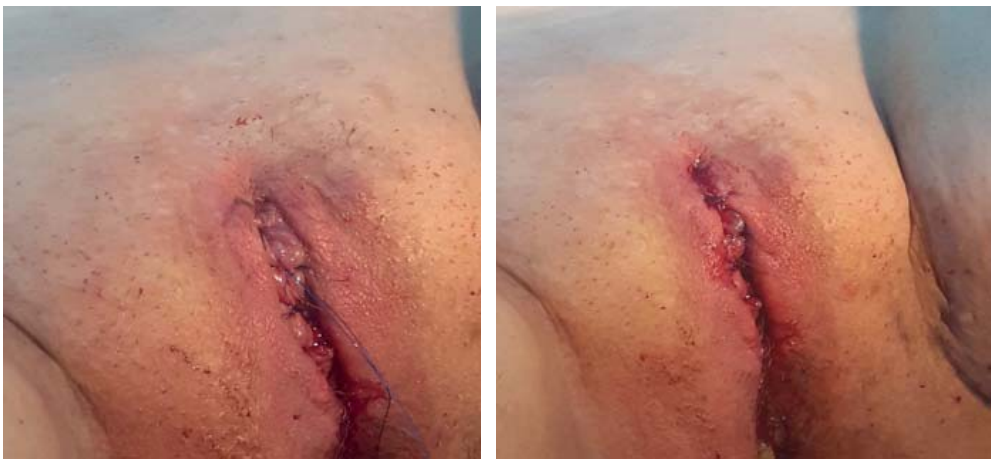

Fig. 3, 4. Reconstructed anatomy and shaped foreskin (left); Clitoroplasty completed (right)

Fig. 5. Status at 6 months after surgery

hyperandrogenism, Prader distinguishes 5 stages in girls:

1. Clitoral hypertrophy, normal vaginal entrance.

2. Clitoral hypertrophy, narrowed vaginal entrance due to partial fusion of the labia minora.

3. Emphasized clitoral hypertrophy, persistent urogenital sinus is observed.

4. The hypertrophied clitoris resembles a penis and the labia majora imitates the scrotum, the vagina opens into the urethra and at the base of the clitoris.

5. Male external genitalia, possibly with hypospadias, without testes in the scrotum.

Endocrinopathies, virilizing tumors, and androgen use are the most common hormonal causes of clitoromegaly.

The most common cause of clitoromegaly is congenital hyperplasia of the adrenal cortex [3]. Adrenal hyperplasia due to 21-hydroxylase deficiency is a rare, congenital disorder presenting with hyperplasia of the adrenal glands in utero due to lack of cortisol due to 21-hydroxylase enzyme deficiency associated with hyperandrogenism. Severe cases occur in 1 in 10-14,000 newborns. Mild cases without classic symptoms are much more common, 1:1000.

Hormone-producing ovarian tumors that can cause clitoromegaly are androblastomas. They originate from the Sertoli-Leydig cells of the hilus of the ovary. They produce androgens that lead to menstrual disorders – amenorrhea and to virilization of women - enlargement of the clitoris and increased hair growth. Very rarely, an androblastoma is hormonally inactive or secretes estrogens.

Non-tumor causes of clitoromegaly can be neurofibromatosis and epidermoid cysts of the clitoris.

Neurofibromatosis (benign) or the so-called von Recklinghausen's disease has two separate, genetically and clinically different forms – type 1 and type 2. The disease is characterized by a wide variability of the clinical picture – from mild asymptomatic forms to very severe manifestations of the clinical picture and a high risk of life-threatening malignant diseases of the patient. Type 1 is more common (1:3,000 – 1:4,000). It is inherited in an autosomal dominant manner. Women suffering from this disease develop benign and malignant tumors of the central and peripheral nervous system. In girls, it can rarely lead to enlargement of the clitoris and labia [4]. Epidermoid cysts of the clitoris are one of the rare non-hormonal causes of clitoromegaly. In such cases, a mobile, soft, non-fluctuating mass protrudes from the clitoral region. There is often evidence of trauma in this area in the woman's history [5].

Clitoroplasty is the method of choice in the treatment of clitoromegaly. It aims to improve the psycho-

emotional state of the woman. At the same time, it is necessary to preserve sexual function and sensitivity with a mandatory cosmetic effect.

For the first time, Young performed a similar plastic correction on a child with congenital adrenal hyperplasia in 1937 [6]. Later J.G. Hampson and J. Money suggest in similar cases to amputate the clitoris [7]. Various operative methods have been described, but the cases of operative correction in sexually mature women in which the dorsal and ventricular vascular-nerve bundles are preserved are few [8]. Clitoroplasty with preservation of neurovascular pedicles, which was performed in the described case, is an optimal volume of surgery in women with clitoromegaly of first degree. In this case, correction of the labia minora and plastic surgery of the introitus vagina is not necessary. Some authors also suggest resection of the corpora cavernosa of the clitoris [6]. In this case, correction of the labia minora and plastic surgery of the introitus vagina is not necessary.

Disclosure summary: *The authors have nothing to disclose.*

Acknowledgements: *This work was performed with support of a research project of Trakia University – Stara Zagora, № BG-RRP-2.004-0006.*

REFERENCES

1. Iezzi ML, Lasorella S, Varriale G et al. Clitoromegaly in Childhood and Adolescence: Behind One Clinical Sign, a Clinical Sea. Sexual Development. 2018; 12(4):163-175.
2. Andrea Prader. 6-6 Delayted adolescence. Clinics in Endocrinology and Metabolism. 1975; 4(1):143-155.
3. Mustafa R, Haleema A, Shafaat U. Congenital adrenal hyperplasia causing clitoromegaly. J Coll Physicians Surg Pak. 2008; 18(6): 378-379.
4. Karac R, Bayne C, Shenoy A, et al. Genital neurofibromatosis presenting as painful clitoromegaly. Urology, 2019; 133, 219-221.
5. Kulaksiz S, Sekerci C. A rare cause of clitoromegaly: epidermoid cyst. Archivio Italiano di Urologia e Andrologia. 2019; 91(2).
6. Young H. Genital abnormalities, hermaphroditism and related adrenal diseases. Baltimore: Williams & Wilkins, 1937.
7. Hampson J.G. Hermaphroditic genital appearance, rearing and eroticism in hyperadrenocorticism. Bull Johns Hopkins Hosp. 1955; 96 (6): 265-73.
8. Uddhav AP, Manasi SD, Shreyasi UP. Reduction Clitoroplasty by Ventral Approach: Technical Refinement. Obstet Gynaecol India. 2019; 69 (1):48-52.