

Case Report

Subtunical Reduction Clitoroplasty of a 22 years old Female for Clitoromegaly with Congenital Adrenal Hyperplasia-Case Report

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Abstract:

Clitoromegaly is an abnormal condition commonly related to congenital adrenal hyperplasia. We presented a 22 years old adult female patient with chief complaint of enlarged clitoris since birth, other female secondary sexual characteristics were normal. The karyotype test result was 46 XX. Subtunical resection of erectile cavernous tissue of the enlarged clitoris with preservation of intact blood supply to the glans is a safe method of diminishing shaft and glans size and allow the patient to do sexual activity after marriage during a two-years follow up.

Keywords: Clitoromegaly, Clitoroplasty.

Introduction:

Congenital adrenal hyperplasia (CAH) is the most common cause of ambiguous genitalia in newborns with an incidence (based on newborn screening programs) of approximately 1:14,000 live births. This condition is inherited as an autosomal recessive disorder and is associated in 75% of patients with a life-threatening salt-losing metabolic condition. 46XX female fetus exposed to adrenal androgens results in varying degrees of virilization of the external genitalia and the distal vagina, although Müllerian precursors of the internal genitalia, fallopian tubes, uterus and proximal vagina develop normally in the absence of Müllerian inhibiting substance¹.

Clitoromegaly is abnormal condition with enlarged clitoris. A study mentioned that a clitoral index (the product of the glans width and glands length) of more than 35 mm² is considered abnormal. Clitoromegaly can be seen since birth, or it can occur later in life. The most common anomaly presenting with clitoromegaly is

CAH, but other pathologic condition can present this appearance, such as Fraser syndrome (an autosomal recessive congenital disorder), polycystic ovarian syndrome, pathological disorders of the ovaries and adrenal, abuse of anabolic steroids, cyst or hemangiomas of the clitoris, or idiopathic^{2,3}. Adult female presenting with clitoromegaly could be psycho-socially distress. Many of them reach sexually active age concern about their sexual life, marriage, and the possibility of having offspring. Ambiguous genitalia represent one of the major challenges for physicians and surgeons⁴. The treatment is multidisciplinary and coordination of neonatology, endocrinology and plastic surgery is essential for an acceptable treatment⁵.

Clitoroplasty is a term for the surgical creation of a clitoris, in transsexual and transgender women (as part of sex reassignment surgery), or restoration in the case of procedures reversing the damage caused by female genital cutting. Women and girls with Congenital

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Adrenal Hyperplasia may also undergo to correct clitoral size in accordance to the female anatomy or complications of the conditions⁶. Modern day treatment has evolved from clitorrectomy to techniques of

recession and resection to reduce clitoral size, each with many variations. Resection techniques involve mobilization of the shaft with relocation under the pubis or in the subcutaneous tissues. Resection may be done by either total or subtotal removal of corporeal bodies and surrounding tunica albuginea or by subtunical resection of the erectile tissue only.

The goals of clitoroplasty are to reduce the size of the hypertrophied clitoral shaft and glans while preserving normal glans sensation and creating normal glans appearance. This goals may be accomplished by recession and excisional resection techniques, preserving glans and dorsal neurovascular bundle in each instance⁷.

Case Report: A 22 years old female presented with facial hair and enlarged clitoris. The clitoris was enlarged since birth, but the patient and the family did not seek any treatment. The patient had menarche at age 13 and got regular periods since then. The patient had no gender dysphoria and keen to get married. She was disturbed because her clitoris could be enlarged and tumescence. There were no abnormalities for urination and defecation. There was no suspicious of disorder of sex differentiation in the family history.

On physical examination, the phallus measured 7 cm in length and 2.5 cm in width, a single and proximal located orifice, and the labial fold normal. There was no palpable gonad nor any virilizing sign. Other appearances of female secondary sexual features were normal. Further evaluation of the patient did not find other gynaecological or systemic disorders.

Abdominal ultrasonography revealed no abnormality, USG of TVS done with high resolution probe revealed uterus was normal in size, anteverted and echogenically uniform. No focal lesion was seen. Endometrial thickness was normal, both the ovaries were normal in size, one mature follicular cyst measured about 17x15 mm in size in left ovary. Hormone assay revealed Serum Testosterone 340.5 ng/dl (above normal level), Prolactin 29.9 ng/ml (above normal), FSH 2.67 mIU/ml (within normal limit), LH 2.20 mIU/ml (within normal limit), TSH 2.36 micro IU/ml (within normal limit), Total T4 8.16 microg/l (within normal limit) and Random Plasma Glucose 4.3 mmol/l. Serum Electrolytes were within normal limit. Cytogenic report was apparently normal female Karyotype (46,XX). She was diagnosed as clitoromegaly with congenital adrenal hyperplasia. Endocrinologist advised to take oral prednisolone.

Subtunical reduction clitoroplasty was done in the Department of Burn and Plastic Surgery, Bangabandhu Sheikh Mujib Medical College Hospital, Faridpur on August, 2020.

Clitoroplasty Technique: The patient underwent clitoroplasty with preservation of the neurovascular pedicles under general anesthesia. The procedure begun with the patient in the lithotomy position. After inspection of the introitus and perineum, there was separate vaginal and urethral orifice. A traction suture of 4/0 prolene was placed in the glans of clitoris. A small volume of epinephrine (1 in 100,000) was injected at the coronal margin of the glans to hemostasis. An incision was made around the dorsal margin of glans close to corona. It was not carried around the full ventral circumference but was extended inferolaterally into lateral attachment of the prepuce (usually at approximately 4 o'clock and 7 o'clock on the glans). This left the broad midline attachment of the mucosa and urethral plate to the glans intact. The clitoris was completely degloved dorsally, preserving the dorsal neurovascular bundles on the surface of the corpora cavernosa⁸. This dissection was extended proximal to the bifurcation of the corpora. Each crus was isolated lateral to the midline and ligated in situ. Corporeal reduction could be accomplished by subtunical reduction⁹. A longitudinal incision was made on the lateral aspect of one corporeal body through buck's fascia and tunica albuginea. The cavernous tissue was resected using sharp dissection and both corpora could be in one package.

The base of the glans was sutured to the divided corpora with 4/0 vicryl. The ventral aspect of the glans was then sewn to the ventral aspect of the tunica albuginea at the level of the crural bifurcation while the dorsal side was fixed to the pubic bone approximately 1 cm above the original bifurcation of the corpora. The prepuce was incised in the midline to reconstruct the labia minora. The skin was closed with 4/0 vicryl sutures. The drain was removed one day after surgery. The Foley catheter was maintained for one week and the healing process was uneventful. No complication was noticed during follow-up.

During one year follow-up, the functional and aesthetic outcome was good. Three months after surgery, the patient got married and did not have any complaint about the sexual function such as pain or difficulty during intercourse, nor reduced sensitivity of the clitoris.



Fig. 1: (A) Phenotypic appearance of the patient before surgery. (B) Ultrasound of the internal genitalia.



Fig. 2: (A) Peroperative photograph, (B) Postoperative photograph



Fig. 3: Follow up after 2 weeks

Discussion:

Clitoromegaly can be seen since birth or it can occur later in life. Even though the most common condition related to clitoromegaly is CAH caused by an enzyme defect in

the normal pathway of steroid biosynthesis, it may result from a variety of conditions. The causes of clitoromegaly can be classified into hormonal conditions, non-hormonal conditions, pseudoclitoromegaly, and idiopathic. A scrutinized evaluation includes a detailed history and physical examination and further laboratory and radiology evaluation are required to search for the etiology of this condition^{10,11}.

The clitoris is the erectile part of the external genitalia, it has homologous properties with the penis¹². The major difference between female external genitalia and male genitalia is that female genitalia are separate from the urethra.

Surgical reconstruction of ambiguous external genitalia in female (feminizing genitoplasty) may include clitoroplasty, vaginoplasty and labioplasty. In presence of idiopathic or isolated clitoromegaly, clitoroplasty may alone suffice. In most situations, however, clitoroplasty, the mainstay of feminizing genitoplasty, must be planned and carried out in concert with labioplasty and simultaneous or delayed vaginoplasty.

The objectives of clitoroplasty are preservation of sexual arousal function, sensation and cosmesis. Historically, until 1960's, clitoral hypertrophy was treated by clitorectomy^{8,13}. Surgical method of correction of clitoral hypertrophy was first described in 1934 by Young, who performed an operation for clitoral reduction in a child with CAH¹⁴. Several clitoroplasty methods have been reported, but few describe preservation of dorsal and ventral neurovascular bundles in sexually mature women. Clitoroplasty with preservation of the neurovascular pedicle may be the optimal operative technique for the treatment of clitoromegaly.

Conclusion:

The goals of clitoroplasty are feminization, preservation of function and sensation, and cosmesis. To achieve these goals, the surgeon diminish clitoral length and diameter, decrease the size of glans clitoridis, preserve clitoral blood supply and sensation, and coordinate clitoroplasty with simultaneous or delayed vaginal /labial reconstruction.

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