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Idiopathic Isolated Clitoromegaly: A Report of Two Cases

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Abstract

Background

Clitoromegaly is a frequent congenital malformation, but acquired clitoral enlargement can be rarely detected.

Methods

Two cases of clitoromegaly treated in Ataturk Training Hospital will be presented in this report.

Results

Histories obtained from the two patients revealed that the clitoris was gradually enlarged for the last three years. Neither gynecological nor systemic abnormalities were detected in both patients. Results of karyotype analyses and hormonal tests were normal. Abdominal and gynecological ultrasound did not show any cystic lesion or abnormal finding. CT scan of the adrenal glands was normal. Clitoroplasty with preservation of neurovascular pedicles were performed for the treatment of cliteromegaly.

Conclusion

The patients were diagnosed as “idiopathic, isolated” clitoromegaly. To the best of our knowledge, there has been no detailed report about idiopathic clitoromegaly in the literature.

Case reports

Two cases with clitoromegaly were treated in Ataturk Training Hospital, Izmir, Turkey.

A 22-year-old gravida 0 and 19-year-old gravida 0 presented with adult clitoromegaly, which was emotionally embarrassing. One had a phallus 20 mm in length, which increased by 30 mm with arousal (Figure 1) and the other had a phallus 30 mm in length, which increased by 40 mm with arousal (Figure 2). Secondary sexual features were otherwise normal. Sexual hair was normal and there are no sign of hirsutism. Patients were not obese and their weights were 65 and 68 kg. Neither of the patients had the signs of polycystic ovaries. Histories taken from both patients revealed a gradually growing clitoris in the last three years. Neither gynecological nor systemic abnormalities were detected in both patients. They did not have a history of drug abuse or family history of clitoromegaly. They did not note clitoral irritation secondary to masturbation or other sexual functions. They had just “isolated” clitoromegaly as an abnormal finding on all detailed physical examinations.

Karyotype analysis was done in both cases and reported as 46, XX. Results of routine laboratory tests were normal. In addition, levels of electrolytes, oestradiol, SHBG, testosterone, androstenedione, DHEA-S, FSH, LH, 17-OH-P, prolactin, ACTH, cortisol PL, Deoxycorticosterone, Deoxycortisol 11, T3, T4, TSH, β HCG, CEA were measured before operations and the results were normal. 17-ketosteroid output in 24-hour-urine specimen was normal in both patients. Abdominal and gynecological ultrasound did not show any cystic lesion or abnormal finding. Computed tomography scan of the adrenal glands was normal.

No abnormality suggestive of clitoromegaly was found in all laboratory and radiological tests. Patients were diagnosed as “idiopathic, isolated” clitoromegaly.

Clitoroplasty with preservation of the neurovascular pedicles were planned and the patients were operated under general anesthesia. A traction suture of 3/0 nylon was placed in the glans of clitoris (Figure 3). An incision was made on the lateral phallus perpendicular to the axis of

the clitoral shaft, and carried through a 270 degree semicircular arc to the base of the glans as described by Papageorgiou et al [1]. Two longitudinal incisions were made laterally to the dorsal neurovascular bundle. Two crura were identified, clamped and mid-body of the clitoris was resected. The base of the glans was sutured to the divided corpora with 4/0 vicryl, and proximal and distal ends of corpora were closed with 4/0 vicryl. The skin was closed with 4/0 vicryl sutures as well. Pathological examinations of resected specimens were made and showed “normal corporal tissue”. There was no abnormal finding on microscopic examination of the specimen obtained from clitoral and submucosal tissue.

Patients were followed up for one year after the operations. There was no early or late post-operative complication. Sensation was normal. Patients were satisfied with the aesthetical and functional results.

Discussion

Clitoromegaly is a frequently seen congenital malformation, but acquired clitoral enlargement may be rarely detected [2]. A detailed history and physical examination **are** required for evaluation of clitoral enlargement because clitoromegaly may result from a variety of conditions [3]. **However, clitoromegaly can be classified into four groups based on the causative factors, i.e. clitoromegaly due to hormonal conditions, clitoromegaly due to non-hormonal conditions, pseudoclitoromegaly and idiopathic clitoromegaly (Table 1).**

Among hormonal conditions giving rise to clitoromegaly are endocrinopathies, masculinizing tumors, exposure to the androgens and various syndromes. The most common cause of clitoromegaly is female pseudohermaphroditism secondary to congenital adrenal hyperplasia (CAH) or adrenogenital syndrome, caused by an enzyme defect in the normal pathway of steroid biosynthesis [4]. Virilization of the external genitalia may cause profound clitoromegaly but rarely causes formation of a true penile urethra. However, clitoromegaly may be accompanied by fusion of the labioscrotal folds and perineoscrotal hypospadias, and the urogenital sinus may persist, so that the vagina does not open to the outside [5].

Tumors are other important factors in the pathogenesis of clitoromegaly. Bilateral hilus cell tumors of the ovary, steroid producing gonadal tumors, adrenal androgen-secreting carcinoma, Leydig cell tumor of the ovaries, metastatic carcinosarcoma of the urinary bladder **have been reported to cause** clitoromegaly [6-9].

Exposure to **androgens** is an important cause of clitoromegaly. An interesting case report was presented by Akcam and Topaloglu [10]. They presented an immature case of clitoromegaly secondary to a blood transfusion from an adult. Fetal exposure to danazol may **also** cause clitoromegaly [11].

Among the non-hormonal conditions leading to clitoromagaly are neurofibroma, epidermoid cysts, various syndromes and nevus lipomatous cutaneous superficialis.

Neurofibromatosis (NF) was one of the most frequently reported reasons of clitoromegaly [12]. The majority of clitoromegaly cases related to NF are congenital.

Sometimes clitoral **cysts** could be **misdiagnosed** as clitoromegaly [3]. They arise from epidermis displaced into the dermis or into the subcutaneous tissue either in prenatal period or after a trauma.

Various syndromes resulting from non-hormonal conditions may also cause clitoromegaly. Kazlauskaite et al reported a case presenting with generalized fat loss, prominent musculature, hepatomegaly, clitoromegaly, mild hirsutism and diagnosed as congenital generalized lipodystrophy (CGL) [13]. CGL is an autosomal recessive disorder, characterized by severe metabolic derangement associated with the absence of subcutaneous adipose tissue, and causes clitoromegaly. Fraser syndrome is another rare reason of clitoromegaly [14]. Turner syndrome (TS) is one of the most common chromosomal disorders in females and results from a partial or complete loss of an X chromosome. Abnormalities include short stature and gonadal dysgenesis. Haddad et al presented a case of clitoromegaly and TS [15]. Androgen insensitivity syndrome is a heterogeneous disorder with a wide spectrum of phenotypic abnormalities, ranging from a complete female to ambiguous forms that more closely resemble males. The primary abnormality is a defective androgen receptor protein due to a mutation of the androgen receptor gene.

Nevus lipomatous cutaneous superficialis (NLCS) is a relatively rare condition characterized histologically by groups of ectopic fat cells dispersed [16] **and it** may cause clitoromegaly when located on the clitoris.

Another type of **cliteromegaly is** pseudohypertrophy of the clitoris **and** often seen in small girls due to masturbation: Manipulations with the skin of prepuce represents repeating mechanical insult, which expands **the** prepuce and labia minora, thus imitating true clitoral enlargement [2].

Several authors **claimed that** the clitoris **was** an erotically important sensory organ worth saving. The goals of clitoroplasty are feminization, preservation of function and sensation, and cosmesis. Historically, until 1960s, clitoral hypertrophy was dealt with surgically by amputation clitoridectomy [4]. Surgical methods for correction of clitoral hypertrophy were first described in 1934 by Young, who performed an operation for clitoral reduction in a child with CAH [17]. 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 is the best operative technique for the treatment of clitoromegaly.

Both patients presented here were diagnosed with “idiopathic clitoromegaly as there was no **history of** drugs and irritation. In fact, they were evaluated according to the criteria on clitoromegaly **reported in the literature**. However, they were not found to have any other illnesses or syndromes **except clitoromagaly**. To our knowledge, there is no detailed report about idiopathic clitoromegaly in the literature. **We performed** clitoroplasty with preservation of neurovascular pedicles **for the treatment of clitoromegaly**.

Competing interests

None declared

Authors' contributions

EC conceived the study and prepared the manuscript draft for submission. **AA, NS, OC** and

YO did the literature search and participated in the preparation of the manuscript.

All authors read and approved the final manuscript.

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Figures

Figure 1 - View of the Case 1.

Figure 2 - View of the Case 2.

Figure 3 - Traction of the clitoris per-operatively.

Tables

Table 1 - Classification of the clitoromegaly based on causative factors

Causative factors of clitoromegaly

A. Hormonal conditions

1. Endocrinopathies
2. Masculinizing tumors
3. Exposure to the androgens
4. Syndromes

B. Non-Hormonal conditions

1. Neurofibromatosis
2. Epidermoid cysts
3. Syndromes
4. Nevus

C. Pseudoclitoromegaly

D. Idiopathic