

Idiopathic isolated clitoromegaly: Report of two cases

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Abstract

Background

Clitoromegaly is usually seen congenital malformation but rarely acquired clitoral enlargement can be detected

Methods

Two cases with clitoromegaly who were treated in Ataturk Training Hospital were presented in this study.

Results

They described gradually growing of their clitoris within last three years. Neither gynecological nor other systemically abnormalities were detected in both patients. Karyotype analysis and hormonal test results were normal. The 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 in the treatment of the patients.

Conclusion

Our patients were diagnosed as “idiopathic, isolated” clitoromegaly. Best of our knowledge, there is no detailed report about the idiopathic clitoromegaly in the literature.

Case report

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

22-year-old gravida 0 and 19-year-old gravida 0 presented adult clitoromegaly that was emotionally embarrassing. First case had phallus was 20 mm long and increased 30 mm with arousal (figure 1) and second case had phallus was 30 mm long and increased 40 mm with arousal (figure 2). Secondary sexual features were otherwise normal. Sexual hair was normal with no hirsutism and obesity in both cases. They described gradually growing of their clitoris within last three years. Neither gynecological nor other systemically abnormalities were detected in both patients. They had no drug or family history. They did not describe clitoral irritation secondary to masturbation or other sexual functions. They had just “isolated” clitoromegaly as abnormal finding in all detailed physical examinations.

Karyotype analysis was done in both cases and reported as 46, XX. Results of routine laboratory tests were normal. Also, preoperatively, levels of electrolytes, oestradiol, SHBG, testosterone, androstendione, DHEA-S, FSH, LH, 17-OH-P, prolactine, ACTH, cortisol PL, Deoxycorticosterone, Deoxycortisol 11, T3, T4, TSH, β HCG, CEA were calculated and results were normal. Output of the 17-ketosteroid in 24 hour urine specimen was normal in both patients. The abdominal and gynecological ultrasound did not show any cystic lesion or abnormal finding. CT scan of the adrenal glands was normal.

No abnormality which could be explained the clitoromegaly was found in all laboratory and radiological tests. Patients were diagnosed as “idiopathic, isolated” clitoromegaly.

Clitoroplasty with preservation of neurovascular pedicles were planned and 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 axia of 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. Skin was closed with 4/0 vicryl sutures also. Resected specimens were evaluated pathological and reported as “normal corpora tissue”. There are no abnormal microscopical findings in the specimen from clitoral and submucosal tissue.

Patients were followed up one year post-operatively. There was no early or late post-operative complication. Sensation was normal. Patients were satisfied aesthetical, functional and emotional.

Discussion

Clitoromegaly is usually seen congenital malformation but rarely acquired clitoral enlargement can be detected [2]. A detailed history and physical examination is required for evaluation of clitoral enlargement, because clitoromegaly can result from a variety of conditions [3]. The most common cause of the clitoromegaly is female pseudohermaphroditism secondary to congenital adrenal hyperplasia (CAH, adrenogenital syndrome), caused by an enzyme defect in the normal pathway of steroid biosynthesis [4]. The result is overproduction of androgenic steroids and masculinization of the external structures, which, because they contain 5 α -reductase, are readily virilized [4]. Congenital adrenal hyperplasia (CAH) is an autosomal recessive disorder caused by a defect in any of the five enzymatic steps required to synthesize cortisol from cholesterol. The most common form is 21-hydroxylase (21-OH) deficiency which accounts for 90-95 % of cases with CAH. A further 5-8 % of cases are associated with a deficiency of 11 β -hydroxylase (11 β -OH) and all other enzymatic deficiencies together account for less than 5 % of cases with CAH [5]. Virilization of the external genitalia can 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 [6]. 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 ovary, metastatic carcinosarcoma of the urinary bladder are the reported tumors which caused to clitoromegaly in the literature [7-10].

Exposure to androgens is an important cause of the clitoromegaly. An interesting case report was presented by Akcam and Topaloglu [11]. They presented an immature case with

clitoromegaly secondary to the blood transfusion from adult. Fetal exposure to danazol may cause to clitoromegaly [12].

One of the most reported reason of the clitoromegaly was the neurofibromatosis (NF) [13]. The majority of clitoromegaly cases related with NF are congenital. Sometimes clitoral cysts could be evaluated as clitoromegaly [3]. They arise either from epidermis displaced into the dermis or into the subcutaneous tissue, either prenatal or after trauma.

Some syndromes may cause to clitoromegaly: Kazlauskaite et al presented a case who had generalized fat loss, prominent musculature, hepatomegaly, clitoromegaly, mild hirsutism and diagnosed as congenital generalized lipodystrophy (CGL) [14]. Congenital generalized lipodystrophy (CGL) is an autosomal recessive disorder, characterized by severe metabolic derangement associated with the absence of subcutaneous adipose tissue and caused the clitoromegaly. Fraser syndrome is another rare reason of the clitoromegaly [15]. Turner syndrome (TS) is one of the most common chromosomal disorders in females and results from partial or complete loss of an X chromosome. Abnormalities include short stature and gonadal dysgenesis. Haddad et al were presented a case with clitoromegaly and Turner syndrome [16]. The androgen insensitivity syndrome is a heterogeneous disorder with a wide spectrum of phenotypic abnormalities, ranging from 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 lipomatosus cutaneous superficialis(NLCS) is a relatively rare condition and this clinical condition is characterized histologically by groups of ectopic fat cells dispersed [17]. NLCS may be caused the clitoromegaly when located on the clitoris.

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

Several authors recognized the clitoris as 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 congenital adrenal hyperplasia [18]. Several clitoroplasty methods have been reported, but few describe preservation of dorsal and ventral neurovascular bundles in sexually mature women. Clitoroplasty with preservation of neurovascular pedicle is the best operative technique in the treatment of the clitoromegaly.

Finally, our both patients were evaluated according to the criteria's of studies related with clitoromegaly, presented in the literature. We could not determine any ill or syndrome in these patients. Since there was no drug and irritation history we defined "idiopathic clitoromegaly". Best of our knowledge, there is no detailed report about the idiopathic clitoromegaly in the literature. According to results of our study, clitoromegaly can be classified as listed Table 1.

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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Figure legends:

Figure 1: View of the Case 1.

Figure 2: View of the Case 2.

Figure 3: Traction of the clitoris per-operatively.

Table 1

Classification of the clitoromegaly

Reasons of the clitoromegaly
A. Hormonal reasons
1. Endocrinopathies
2. Masculizing tumors
3. Exposure to the androgens
4. Syndromes
B. Non-Hormonal reasons
1. Neurofibromathosis
2. Epidermoid cysts
3. Syndromes
4. Nevus
C. Pseudoclitoromegaly
D. Idopathic



Figure 1



Figure 2



Figure 3