Case report

CLITOROMEGALY: CASE REPORT AND LITERATURE REVIEW

ABSTRACT

Aim:

To highlight the presence of clitoromegaly in a pregnant woman, with no history of difficulty in achieving pregnancy.

Background: Clitoromegaly is an abnormally large clitoris.

The clitoris develops from the genital tubercle in the female and forms the penis in the male. It could be congenital or acquired. When seen in newborns, its likely cause is Congenital Adrenal Hyperplasia (CAH) and this condition is secondary to enzyme deficiency -21 hydroxylase deficiency or 11Beta hydroxylase deficiency.

Patients with clitoromegaly are usually associated with infertility, because of the high circulating androgens.

Method:

The case report was that of a 37 years old Gravida 4, Para 1^{+2} who was admitted for cervical cerclage insertion due to previous recurrent mid-trimester miscarriages. Clitoromegaly was observed in theatre as an incidental finding.

Results: Investigation results were karyotype 46XX, Normal Uterus, fallopian tubes and ovaries. The cervix was short with funnelling of the Internal Cervical OS.

Cortisol level was low and Testosterone level was minimally high.

ACTH stimulation test noted adrenal hyperplasia.

21 hydroxylase deficiency was Non-Classical CAH. This is a mild form of Congenital Adrenal Hyperplasia. Patients usually may or may not present with symptoms.

CONCLUSION

This is a case presentation of a patient with clitoromegaly, with investigation results showing a mild form of adrenal hyperplasia. Infertility was not a problem, as patients had conceived four times.

The small penis-like structure of her clitoris was also not a problem for her.

Key Word:

Clitoromegaly, Adrenal Hyperplasia, 21- hydroxylase deficiency, Androgens.

Introduction

Clitoromegaly or macro clitoris is an abnormally large clitoris.[1,2,3] The clitoris in females is the equivalent of the penis in males. They both develop from the genital tubercle during organogenesis. 2,5 Theabsence of androgens leads to the development of the clitoris in females as part of the external genitalia.[5]
The presence of androgens causes the genital tubercle to develop into the male penis.
The Clitoris has about 8,000 nerve endings responsible for producing pleasure.Clitoromegaly is a rare Gynecological condition.[1,2]
It can be present at birth (congenital) or develop later in life. Exposure to androgens in a phenotypic female can cause clitoral enlargement.
Pseudo-clitoromegaly can be found in girls as a result of constant masturbation.[7]
Clitoromegaly can also be idiopathic.
In adults, dimensional criteria are, according to Brodie, a minimum length hood And widths of 27.4mm and 8mm.
In Clitoromegaly, these dimensions are exceeded.[1,2,4]
Female patients with Congenital Adrenal hyperplasia, an autosomal recessive disorder, may have impaired cortisol synthesis due to enzyme deficiencies e.g. 21 hydroxylase deficiency. They produce androgens in utero, thereby undergoing varying degrees of virilization of the external genitalia, which can result in some cases, the development of the clitoris as a small penis.[4,5,6] This was the incidental finding in the case presented. Complaints of clitoral enlargement may present later in life due to acquired conditions that cause excess androgen secretion. These conditions are associated with infertility. In this case report, the presenting

complaint was not Clitoromegaly. It was an incidental finding on examination of the patient.

AIM/OBJECTIVE: To highlight a case of clitoromegaly, with laboratory confirmation of Non-Classical Congenital adrenal hyperplasia (N-C CAH) with no history of infertility

Case Report

This was a case report of Mrs R.A, a 37year old booked Gravida 4, Para 1+2 who presented to our facility at 15 weeks gestation with a history of two recurrent mid-trimester miscarriages.
She was admitted through the Gynaecological clinic for a prophylactic cervical cerclage insertion.
She was a Known Diabetic before pregnancy and had been on subcutaneous Insulin she was a known hypertensive and was seen regularly by the physician and endocrinologist. No history of exposure to anabolic steroids.
She had her first delivery after 1 year and 8 months of marriage. The baby is alive and well. This was followed by 2 spontaneous miscarriages at 20 weeks and 18 weeks respectively. Thereafter she was counselled to come for a prophylactic cervical cerclage insertion in her next pregnancy. She was presently 15 weeks pregnant and wanted cerclage insertion.
On Examination,
She was a young lady in no obvious distress, her voice was not deep, and her body mass index was 32kg/m2.
Her chest was clinically clear, pulse rate was 92 b/min and blood pressure was 140/95 mmHg
Her abdomen was enlarged with male pattern hair distribution. The fundal height was 14/52. Her liver, spleen and kidneys were not palpable. Her packed cell volume was 33% and white blood cells were within normal.Her Urinalysis result was also normal. An Obstetric ultrasound scan noted a viable intra-uterine pregnancy at 15 weeks gestation. The cervical length was <2.5cm.She was counselled about her condition and the procedure to be done. She gave written consent.
After pre-operative preparation, she was taken to the theatre for the procedure. An enlarged clitoris was noted, with the clitoris appearing like a small penis (as seen in the photograph). Cervical cerclage with Marceline tape was inserted using McDonald's method. Procedures were well tolerated and she was observed in the ward for 72 hours. The patient was counselled about her clitoromegaly.

She gave verbal consent to carry out more investigation but refused any treatment for clitoromegaly. She said that her husband and herself, were satisfied with the size of her clitoris, thus no need for treatment.

Further investigations done noted karyotype 46, XX, ACTH stimulation test noted adrenal hyperplasia, 21 hydroxylase deficiency, cortisol level was 130n mol/L
Total Testosterone level was 70ng/dl
Her post-operative treatment included analgesics, and prophylactic antibiotics and to continue with her anti-hypertensives and insulin

Her pregnancy was uneventful. She came for her routine antenatal visits. At 37 weeks gestation, her cervical cerclage was removed. 48 hours after removal of cerclage, labour pains commenced. This progressed to the delivery of a live male baby weighing 2.8kg. There were no complications. She was discharged home, to see her physician and endocrinologist in the outpatient clinic, she is yet to come for her 6weeks post-natal visit.

Fig.1.Morphological view of Clitoromegaly

DISCUSSION

Clitoromegaly is a rare condition.[2] Females that have clitoromegaly usually present with infertility.[2,5] In the case presented there was no history of difficulty in achieving pregnancy. When clitoromegalyis detected at birth in a female baby, the likely cause is 21-hydroxylase deficiency due to congenital adrenal hyperplasia. Another enzyme that may be deficient is 17 Beta hydroxylase.[4,5] These enzyme deficienciescause a decrease in cortisol production and an increase in circulating androgens. The androgens cause virilization of the female fetus. The circulating androgens can cause the clitoris to appear like a small penis, as seen in our patients (see photography)

She had hirsutism. Her body mass index was 32kg/m2 (obesity). There was a positive history of menstrual irregularities (Oligomenorrhea). She had been a known hypertensive and Diabetic five years ago. She has been managed by physicians on anti-hypertensives and subcutaneous insulin.

Her first pregnancy was conceived spontaneously after marriage and was carried to 36 weeks gestation when she had a spontaneous vaginal delivery. This was followed by two mid-trimester miscarriages at 20 weeks and 18 weeks respectively. Her doctor advised her to have a prophylactic cerclage inserted in her next pregnancy.

This was not the typical presentation of patients with clitoromegaly. The circulating androgens prevent ovulation, thus making conception difficult. From her investigation results, she had a mild form of 21-hydroxylase deficiency. In mild cases, patients may or may not have symptoms.[4,5,7] The interesting finding, in this case, is the clitoromegaly appearing like a small penis.

Her Cortisol level was low Testosterone was marginally high. This was the reason for her hirsutism and clitoromegaly. Co-management with the physicians helped to ensure that her hormones were balanced and her blood pressure controlled. This resulted in satisfactory outcomes for the mother and baby.

Treatment for clitoromegaly could be medical or surgical. Medical treatment is to reduce androgen levels or to block androgen receptors.

The surgical treatment isclitoroplasty, – reducing the size of the clitoris, with conservation of nerve supply.

This patient refused any form of treatment for her Clitoromegaly and said that she and her spouse are satisfied with the size.

LITERATURE REVIEW

Clitoromegaly or Macro Clitoris is an abnormally large clitoris, with dimensionsgreater than the accepted 27.4mm and 8mm hood length and width,in adult females[3].
It can be congenital or acquired. Incidence is 1:10,000- 1: 20,000 females.[1,2]
The clitoris in the female is the equivalent of the male penis. They both develop from the genital tubercle. The presence of androgens progresses to the formation of the penis. The absence of androgens leads to the formation of the clitoris.[3,4]Exposure of the female fetus(Karyotype 46 XX) to excess androgens lead to clitoromegaly.
In Congenital Adrenal Hyperplasia, an autosomal recessive disorder, there is impaired cortisol synthesis, and production of androgens in-utero as a result of a deficient enzyme 21 hydroxylase, causing the fetus to undergo varying degrees of virilization of the external genitalia, which in some cases, can result in development of normal penile morphology.
Congenital Adrenal Hyperplasia (CAH) is divided into the classical CAH. This is the severe form of the disorder. It is further subdivided into:
a salt-losing form or b- simple virilizing form.
The non-classical CAH (NC-CAH) is milder and the patient may or may not present with symptoms.[3,4,5,8]
Other conditions that cause hormonal imbalance can also lead to an enlarged clitoris.[8,12]
Examples of these are:

- Polycystic Ovarian Syndrome (PCOS). There is excess androgen production that could lead to virilization.
- Using anabolic steroids.
- The presence of a tumour on the adrenal gland.
- Using Testosterone replacement therapy for low libido.

Other causes of Clitoromegaly that are not related to hormones include: abscess, vulvitis,neurofibromatosis,clitoral leiomyoma and secondary to frequent masturbation.[10,11]
Treatment of Clitromegaly could be medical- Involving the use of hormones, antibiotics or counselling depending on the cause.
Surgical treatment-Clitoroplasly, Reducing the size of the clitoris.[10,11,12] This could lead to complications like infection, bleeding, and decreased sexual pleasure due to desensitization of the clitoris.[10]
Clitromegaly in a pregnant woman, with no history of infertility is significant. Hence, there was no desire for the treatment of her enlarged clitoris.

CONCLUSION

This case study features a patient with clitoromegaly whose tests revealed a minor type of adrenal hyperplasia. Patients had conceived four times, thus infertility was not an issue. She did not find it bothersome that her clitoris had a little penis-like structure.

Ethical consideration:
Being a Teaching hospital, with teaching and research as our focus, ethical approval was given for taking photos and case discussion.

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