International Journal of Basic and Applied Medical Sciences ISSN: 2277-2103 (Online)
An Open Access, Online International Journal Available at http://www.cibtech.org/jms.htm
2014 Vol. 4 (2) May-August, pp. 192-194/Tuteja et al.
Case Report

CONGENITAL IDIOPATHIC ISOLATED CLITOROMEGALY

Neeraj Tuteja, Sudha Saluja, Sanjay K. Jain, Avinash Yadav, L. D. Agarwal and *N. Hooja
SMS Medical College, Jaipur (Rajasthan), India
**Author for Correspondence*

ABSTRACT

Clitoromegaly is a frequent congenital malformation as part of Congenital Adrenal Hyperplasia being, the most common cause, acquired clitoromegaly is relatively rare. We present a case of isolated congenital clitoromegaly, which is a rare entity.

Keywords: *Congenital Idiopathic, Clitoromegaly*

INTRODUCTION

Clitoromegaly (macroclitoris) is an abnormal enlargement of the clitoris.The clitoris may resemble a small penis. The typical clitoris is defined as having a crosswise width of 3 to 4 mm. (0.12 - 0.16 inches) and a lengthwise width of 4 to 5 mm (0.16 - 0.20 inches).When the clitoris size is greater than 35 mm^2 (0.05 $inches^2$), which is almost twice of the normal size it is called clitoromegaly.Clitoromegaly is also defined as a measure of the clitoral index (width × length in mm) more than 15 mm in the new born and more than 21 mm in an adult woman.

It is a frequent congenital malformation as part of Congenital Adrenal Hyperplasia being, the most common cause, acquired clitoromegaly is relatively rare (Horejsi, 1997). We present a case of isolated congenital clitoromegaly, which is a rare entity.

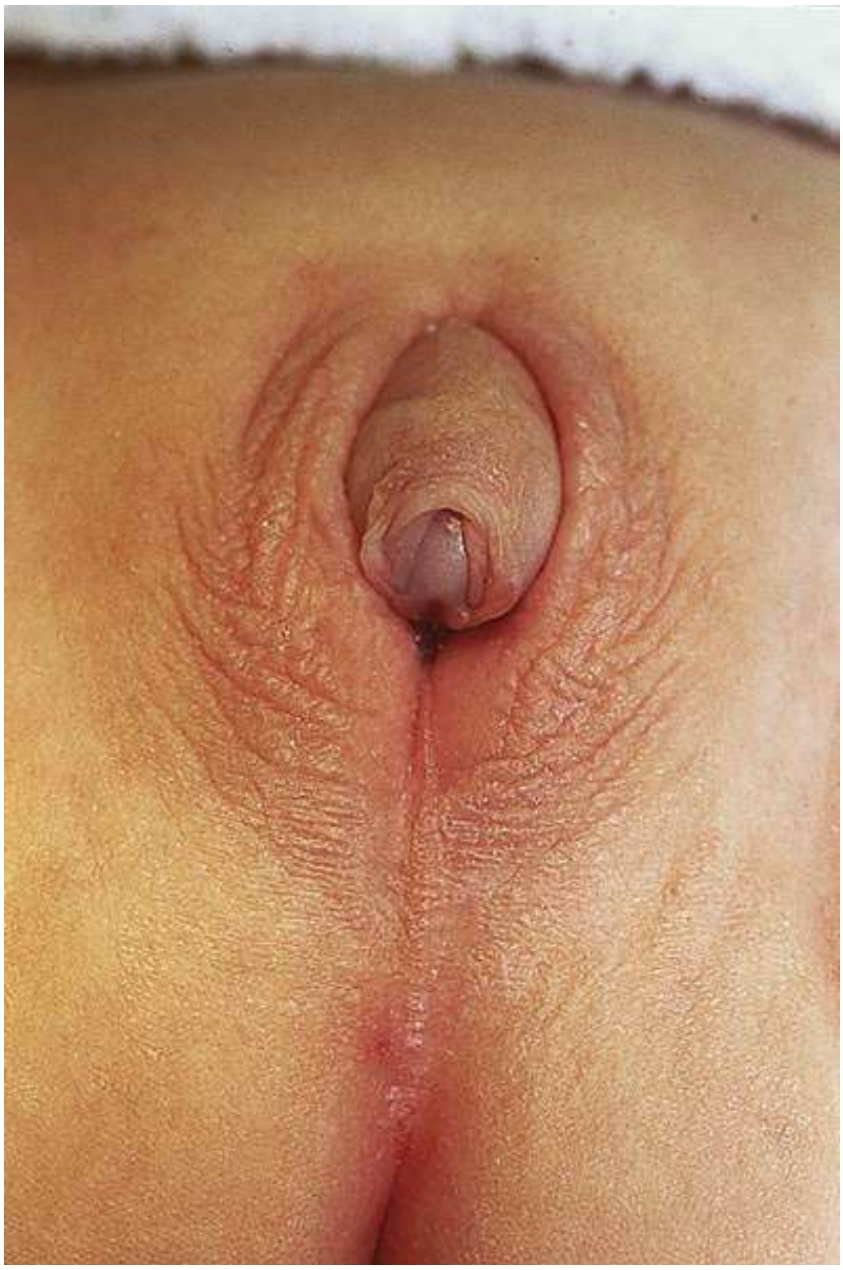
CASES

An 8-year-old girl was brought with complaint of abnormal appearance of external genitalia. The parents noticed enlargement of the clitoris since childhood. Parents were in psychological distress. There was no significant past medical history. Clitoris gradually increases in size. Her mother had not been under hormonal therapy during pregnancy and her 6 year old sister had no anomaly of external genitalia. She was well nourished and averagely built, not obese (17kg) with no pallor/icterus/cyanosis. Her genitalia showed an oval mass in the upper part of urethra that seems to be clitoral mass. There were no pubic hair and pigmentation. The urethral orifice and vaginal orifice existed separately. Anus was normal. There was no virilizing sign or symptom. There was no clinical evidence of adrenal hyperplasia. There was no sign of polycystic ovarian disease.

Laboratory investigations, including complete blood count, electrolytes, coagulation profile. Chromosome analysis revealed 46, XX, and negative for the SRY gene. USG whole abdomen-NAD MRI revealed normal uterus, vagina and ovaries.

Endocrine evaluation showed normal concentrations of adrenocorticotropic hormone (ACTH), follicle stimulating hormone, luteinizing hormone, 17α-hydroxyprogesterone, cortisol, testosterone and dihydroepiandrostenedione. The urinary 17-ketosteroid, 17-hydroxycorticosteroid, pregnanediol and pregnanetriol, were also normal. The concentration of each serum steroid after stimulation by ACTH was normal.

Counseling of the parents and girl was done and clitoroplasty with preservation of neurovascular pedicle was performed. She stood the operation well. On follow up, there was no increase in size of clitoris.



DISCUSSION

Clitoromegaly is a frequently seen congenital malformation, acquired clitoral enlargement is rarely detected the cause of clitoromegaly can be classified into four group. Hormonal condition-Endocrinopathies (Most common cause is secondary to congenital adrenal hyperplasia or adrenogenital syndrome) (Horejsi, 1997), Masculinizing tumors (B/L hilus cell tumor of ovary, Leyding cell tumor), Exposure to androgen (Fetal exposure to danazol), Part of syndromes (Turner syndrome) (Savita *et al.*, 2007). The Non Hormonal conditions may be Neurofibromatosis, Epidermoid cyst, Syndromes, Nevus (Eray *et al.*, 2004). It could be Pseudoclitoromegaly (reported in small girls due to masturbation) or Idiopathic causes.

Treatment includes medical therapy to treat the underlying cause and surgical-clitoroplasty along with preservation of neuro-vascular pedicleas objective is the preservation of sexual arousal function along with cosmetic results (Rangecroft, 2003).

REFERENCES

Eray C, Alper A, Nazan S, Ozgen C and Yucel O (2004). Idiopathic isolated clitoromegaly: A report of two cases. *Reproductive Health* **1** 4.

Horejsi J (1997). Acquired clitoral enlargement. Diagnosis and treatment. *Annals of the New York Academy of Sciences* **816** 369-372.

Inamdar Savita, Inamdar Sameer, Patel Manish and Archana Gupta (2007). Ambiguous genitalia: A spectrum from iatrogenic to genetic disorder, report of two cases. *The Journal of Obstetrics & Ganecology of India* **57**(3) 265-267.

Rangecroft L (2003). Surgical management of ambiguous genitalia. *Archives of Disease in Childhood* **88** 799-801.