International Journal of Research in Medical Sciences
Dhande A et al. Int J Res Med Sci. 2015 Dec;3(12):3887-3888
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20151463

Case Report

Neurofibroma presenting as clitoromegaly

Ashish Dhande, Qurratulain Ali Chougle*, Vikash Lal, Sumeet Gupta, Kirtibala Dhande

Department of Urology, D Y Patil Hospital & Research Centre, Nerul, Navi Mumbai, India

Received: 14 October 2015
Accepted: 20 November 2015

***Correspondence:**
Qurratulain Ali Chougle,
E-mail: qchougle@gmail.com

ABSTRACT

Congenital clitoromegaly is common, but acquired clitoromegaly due to neurofibroma is rare. Here we present a case report of Neurofibroma presenting as clitoromegaly. A 7 year female had history of painless clitoromegaly since 3 years. Her hormonal assays and radiological examination of abdomen and pelvis did not reveal any abnormality. She was subjected to clitoroplasty for correction of clitoromegaly.

Keywords: Clitoromegaly, Intersex, Neurofibromatosis

INTRODUCTION

Neurofibromatosis is an autosomal dominant progressive disorder with an incidence of approximately 1in 3000 live births.[1] Genital lesions are occasionally present, with the vulva being the most common site of involvement. Only rare incidences of clitoral, vaginal, cervical and ovarian neurofibromas have been reported.[2]

CASE REPORT

6 year old girl presented to urology clinic with complaints of painless clitoromegaly since 3 years. Parents were concerned about intersex condition. There was no significant past medical or family history. General examination was unremarkable. Local examination revealed gross clitoromegaly measuring approximately 8-10cm.Vaginal opening showed intact hymen and normal urinary meatus. There was no evidence of any other anomaly. Patients karyotype was 46XX. She was subjected to extensive endocrine and radiological evaluation to rule out precocious puberty and endocrine causes of clitoromegaly. Serum electrolyte, renal function test, thyroid function test and all other routine investigations were sent and found to be within normal limits. 17-hydroxyprogesterone, androstenedione and testosterone levels were also with in normal limits. Abdominal and pelvic ultrasound did not show any abnormality. Patient was planned for elective surgery and resection was carried out with preservation of neurovascular bundle and clitoris. Histopathology of resected specimen showed infiltrating plexiform Neurofibroma. The child was followed up for a period of one year and the follow up did not reveal any recurrence.

Figure 1: Pre op photograph of patient showing hypertrophied phallus like appearance (front).

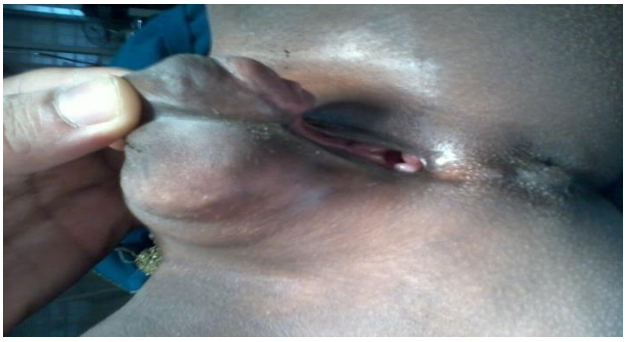

Figure 2: Pre op photograph of patient showing hypertrophied phallus like appearance (lateral).

DISCUSSION

The first description of clitoral neurofibroma was by Haddad and Jones in 1960.[3] In cases of neurofibromas involving the female external genitalia, examination generally reveals clitoral enlargement resembling a phallus. Occasionally, this enlargement masquerades as an intersex disorder and is confused with virilizing congenital adrenal hyperplasia. Of the two pathological subtypes of neurofibromatosis, discrete nodular and plexiform neuromas, the plexiform subtype is more common in urogenital involvement.[4]

After review of this case and the existing literature we conclude that isolated neurofibroma can rarely present as clitoromegaly and create diagnostic dilemma as far as intersex condition is concerned.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Griebel ML, Redman JF, Kemp SF, Elders MJ. Hypertrophy of clitoral hood: presenting sign of neurofibromatosis in female child. Urology. 1991;37:337-9.
2. Jamie AM. Massie, Judith Lacy. Plexiform Neurofibroma Presenting as Clitoromegaly: Case Report and Review of the Literature. 2008; 21(2): 97.
3. Haddad HM, Jones HW Jr. Clitoral enlargement simulating pseudohermaphroditism. AMA J Dis Child. 1960;99:282-7.
4. Kaneti J, Lieberman E, Moshe P, Carmi R. A case of ambiguous genitalia owing to neurofibromatosis-- review of the literature. J Urol. 1988;140:584-5.

Cite this article as: Dhande A, Chougle QA, Lal V, Gupta S, Dhande K. Neurofibroma presenting as clitoromegaly. Int J Res Med Sci. 2015;3:3887-8.