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Clitoromegaly in Neurofibromatosis-A Case Report

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Abstract: *Though clitoromegaly is common,its association with Neurofibromatosis is extremely rare. In this case report a female presented to our OPD with painless clitoromegaly associated with features of Neurofibromatosis 1 for the past 15 years. Her hormonal and radiological profile was normal. Features of Neurofibromatosis were plexiform neurofibroma, cafe au lait macules and axillary freckles.*

Keywords: *Clitoromegaly, Neurofibromatosis, Plexiform neurofibroma, Cafe au lait macules*

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I. Introduction

Neurofibromatosis is an autosomal dominant disorder with an indidence of approximately 1 in 3000 live births.[1] Genital involvement is extremely rare presrenting as clitoromegaly in association with other cutaneous neurofibromas[2].Only rare incidences of clitoral,vaginal,cervical and ovarian neurofibromas have been reported.[3] Patients of Neurofibromatosis-1 may have neurofibromas, cafe au lait spots, axillary freckles,lisch nodulegiant pigmented hairy nevi and sacral hypertrichosis. Neurofibromas result from proliferation of all supporting elements of the nerve fibres.

II. Case Report

A 22 year old female presented to the Dermatology OPD of tertiary care centre of Jharkhand with a plexiform neurofibroma present on the anterior, lateral and medial aspect of arm and forearm, freckles present extensively on almost all parts of the body including axillae, multiple cafe au lait macules in association with

clitoromegaly for the past 15 years. Local examination of the external genitalia revealed gross clitoromegaly measuring approximately 6-7 cms with clitoris resembling phallus. The vaginal opening a urinary meatus were normal. Patient's menstrual cycle was normal in onset, duration and regularity and there was no history of pain in association with clitoromegaly. Patient was adviced elaborate endocrinal and radiological investigations to rule out other causes of clitoromegaly. Her serum electrolyte, serum cortisol and thyroid function test were normal. Her serum testosterone, FSH,LH were also within normal limits and there was no abnormality detected in ultrasonography of abdomen or pelvis.

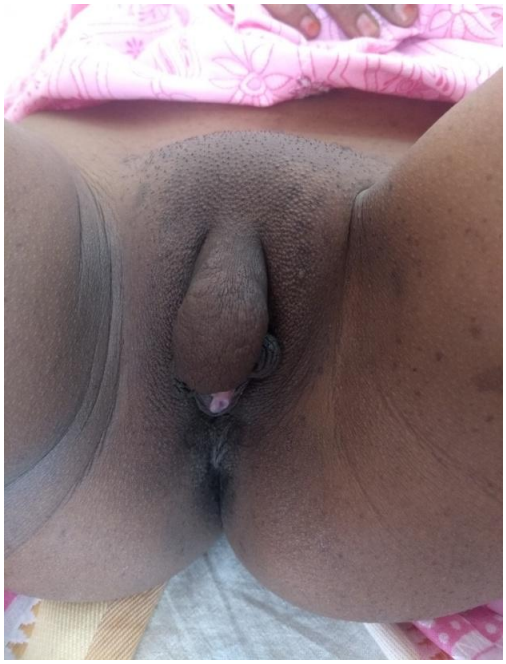

Figure 1. Clitoromegaly resembling phallus

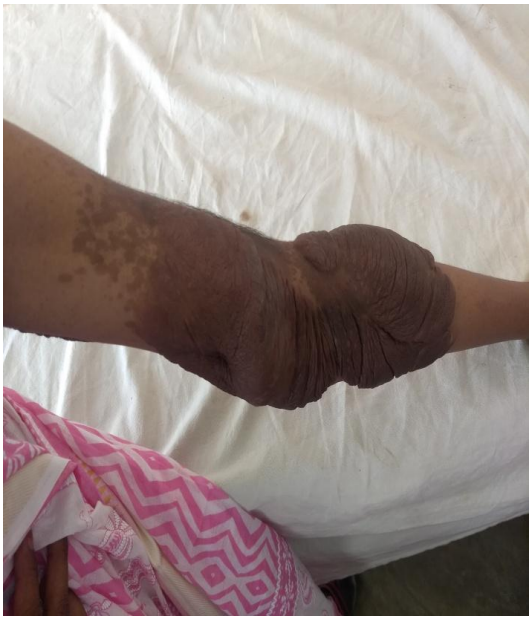

Figure 2. Plexiform neurofibroma

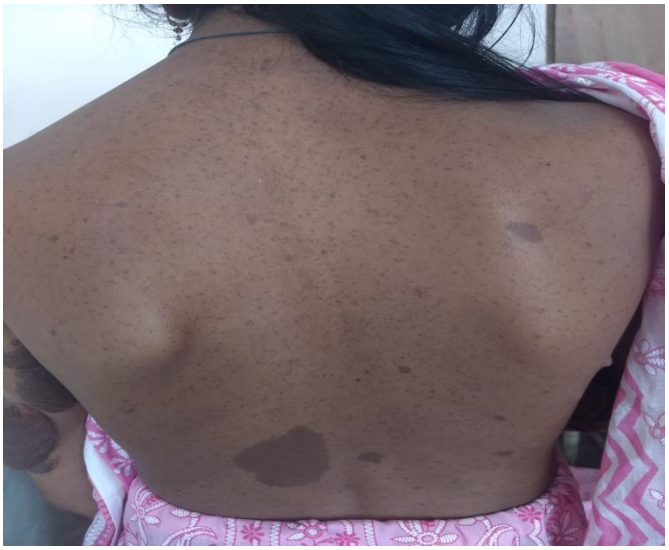

Figure 3. Cafe au lait macules and freckles

III. Discussion

Haddad and Jones gave the first description of clitoral neurofibroma in 1960[4]. Plexiform subtype is more common in urogenital involvement than nodular neurofibroma[5]. Differential diagnosis of clitoromegaly consists of hormonal (ambiguous genitalia,precocious puberty,congenital adrenal hyperplasisa, masculinizing tumours), non hormonal (neurocutaneous syndromes epidermoid cysts,nevus) or could be idiopathic.Current suggested management of clitoromegaly is surgical excision with clitoroplasty.

After review of this case and existing literature we conclude that isolated neurofibroma can rarely present as clitoromegaly.

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