Case report: Clitoroplasty in neurofibromatosis presenting as clitoromegaly

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A B S T R A C T

Genital neurofibromatosis is a rare disorder and clitoral involvement has been reported very infrequently. Clitoromegaly is often presenting sign, in many cases it is congenital. Pathogenesis of clitoral lesion due to neurofibromatosis is similar to other lesion of neurofibromatosis. Occasionally, this enlargement masquerades as an intersex disorder and is confused with virilizing congenital adrenal hyperplasia of the two pathological subtypes of neurofibromatosis.

1. Case report

We present a 5 years old girl with a clitoris resembled as a phallus. The clitoral mass increase in size during the previous 4 years. In all cases diagnosis of clitoromegaly requires basic chromosomal and endocrinologic evaluation. In this patient the chromosomal examination is 46XX, while all endocrinologic evaluation in normal limit, ultrasound for uterus and ovaries and genitogram evaluation also has normal result. Histopathologic examination revealed plexiform neurofibroma.

This patient successfully treated by clitoroplasty, clitoroplasty was performed by preserving the glans of the clitoris and neurovascular bundles (Figs. 1–5).

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Fig. 2. Café au lait spots in neurofibromatosis.

Fig. 3. The gloving of the clitoris.

Fig. 4. Clitoroplasty steps: a) degloving of the clitoris, b) mobilization and separating the neurovascular bundles from the phallus, c) reducing and reconstructing the phallus into the neo clitoris. d) After clitoral reduction and labia majora reconstruction.
2. Discussion

There was not many previous reports of clitoral neurofibromas. The first description of clitoral neurofibroma was by Haddad and Jones in 1960 [1].

A later report by Rink and Mitchell suggested that any child with genital neurofibromatosis be evaluated for bladder neurofibromas based on a few cases with involvement of both external genitalia and bladder [2].

In cases of neurofibromas involving the female external genitalia, examination generally reveals clitoral enlargement resembling a phallus, and some patients report pain if presenting after puberty.

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 [1–4].

Overall, the incidence of malignant degeneration of neurofibromas ranges from 13% to 29%, which increases with age. Thomas and colleagues reported a case of clitoral involvement by malignant schwannoma and described treatment with total surgical removal of the gross tumor [3,4].

After review of this case, we recommend that management of a clitoral neurofibroma consist of excision with all attempts to preserve the clitoris and its adjacent neurovascular structures. We also recommend regular postoperative monitor for local recurrence every 3–6 months of evaluation.

References


Fig. 5. Post operation picture.