Case Report

Isolated Vascular Malformation of the Clitoris: An Uncommon Cause of Clitoromegaly

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Abstract

Clitoral enlargement (clitoromegaly) due to non-hormonal causes, particularly due to vascular malformation, is rare. Only eight such cases are reported in the literature. We report an additional case of isolated vascular malformation implicating the clitoris of a young girl child and its surgical management.

Keywords: Clitoral index, clitoromegaly, hyperandrogenism, vascular malformation

Introduction

Clitoral enlargement (clitoromegaly) is defined as clitoral index (width × length in mm of glans clitoridis) more than 15 mm^2–21 mm^2 in newborn and adult women.[1] It is usually due to hyperandrogenism, i.e., congenital adrenal hyperplasia (CAH).[2] However, occasionally, it is due to preputial or clitoral masses (tumours), clitoral abscess and haematoma. We report a case of an isolated vascular malformation involving the clitoris in a young girl, discuss management and review the relevant literature. Only eight such cases are reported in English literature.

Case Report

A 3.6-year-old healthy female was brought by parents for evaluation of a progressively enlarging mass involving the clitoris since the age of 3 months. She was erroneously investigated for CAH. However, endocrinology was normal, and karyotyping was 46 XX. Ultrasound with colour Doppler revealed a non-homogenous, hypoechoic, vascular lesion at the clitoris; however, other urogenital organs were appropriate for age. The patient was operated under general anaesthesia. Intraoperative examination revealed a dark-coloured, mixed consistency, 15 mm × 25 mm lesion (clitoral index = 375 mm^2) around the clitoris. Urethral meatus was not involved [Figure 1a]. The lesion was meticulously separated from the clitoris, and it was preserved [Figure 1b]. The patient had an uneventful post-operative course, and she was discharged on the 3rd post-operative day. Histopathological

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Table 1: Causes of clitoromegaly

Congenital (CAH)	Acquired	
	Hormonal	Non-hormonal
Classical ambiguous genitalia	CAH	Neurofibromatosis
In syndromic or isolated conditions	Late-onset adrenal or ovarian (androgen secreting) tumours	Epidermoid cyst (in case of trauma caused by genitalia mutilation or spontaneous)
	Iatrogenic androgen exposure	Clitoral or preputial haemangioma/ vascular malformation
		Rhabdomyosarcoma
		Metastatic infiltration
		Clitoral abscess
		Idiopathic

CAH: Congenital adrenal hyperplasia

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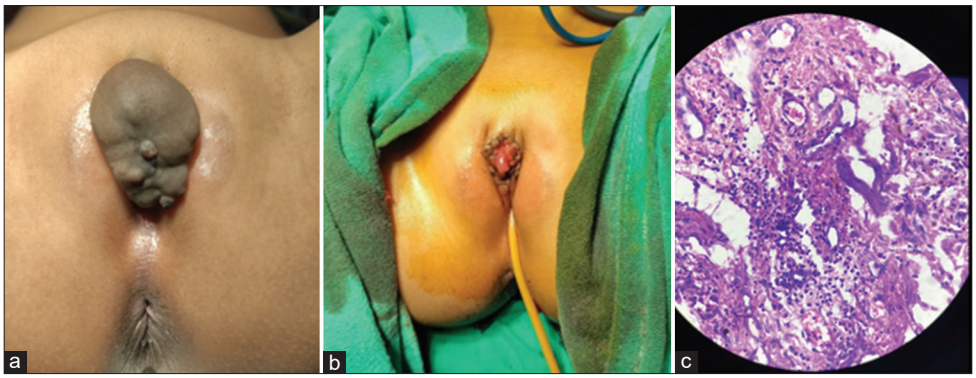


Figure 1: (a) Pre-operative, (b) Post-operative, (c) Histopathological pictures

Table 2: Summary of reported cases of clitoromegaly secondary to vascular lesions

Year	Authors	Clinical features	Outcome/comments
1978	Kaufman-Friedman *et al*.	Isolated haemangioma of the clitoris in an 18-year-old female	Underwent initial inguinal exploration as a case of Intersex; diagnosed during 2nd surgery when uncontrolled bleeding was seen locally and total enlarged mass was excised
1991	Ishizu *et al*.	Large haemangioma involving only the clitoral prepuce in a 4-year-old female; rest of the genitalia and clitoris normal; had earlier had surgery for a large femoral tumour which was also arteriovenous haemangioma	Excision done; arteriovenous haemangioma confirmed on HPE
2006	Haritharan *et al.*	Isolated vascular malformation of the clitoris in a 5-year-old female with enlarged clitoris and history of recurrent bleeding	Delineated on USG and MRI; lesion excised separately from the clitoris and reduction clitoroplasty performed; confirmed on HPE
2009	Bruni *et al*.	Haemangioma of the clitoris presenting as massive clitoromegaly in a 20-year-old female	Diagnosed on HPE
2012	Geramizadeh *et al*.	Clitoromegaly due to cavernous haemangioma in a 16-year-old female	Excised separate from the clitoris; confirmed on HPE
2014	Nayyar *et al*.	Clitoral cavernous haemangioma in a 4-year-old female presenting as clitoromegaly	Excised separate from the clitoris; confirmed on HPE
2018	Barral *et al*.	Cliteromegaly due to venous malformation in a 23-year-old female; also had venous malformations over the craniofacial region	Confirmed on imaging; sclerotherapy instituted
2022	Thakur *et al*.	Clitoral haemangioma in a 4-month-old female. Similar lesions in the perineum, anus and vagina	Excision of clitoral haemangioma followed by oral propranolol for other lesions; confirmed by HPE
2023	Current case	Clitoromegaly due to vascular malformation in a 3.6-year-old female	Excision done separate from the clitoris; confirmed on HPE

HPE: Histopathological examination, USG: Ultrasonography, MRI: Magnetic resonance imaging

examination of the lesion revealed features consistent with a benign vascular lesion [Figure 1c]. At 1-year follow-up, she was asymptomatic with near-normal cosmetic appearance.

Discussion

Clitoromegaly is usually due to raised androgen level [Table 1] as in cases of CAH. Non-hormonal causes are rare, and only a few cases are reported due to neurofibromatosis, epididymal cyst, abscess, haematoma, rhabdomyosarcoma[1,3] and metastasis from bladder carcinoma.[4] Clitoral vascular malformation and haemangiomas are infrequent; only eight such cases have been reported in the literature. Table 2 presents a summary of these cases.[1,2,3,5-9] Although the differences between vascular malformation and haemangiomas are well established now, they have been presented together because earlier this differentiation was not clear. However, it is now known that vascular malformations are pathologic lesions of vascular endothelium, and contrary to haemangiomas, they are present since birth, do not exhibit typical phases of growth and involution and can attain large sizes.[10] Unlike haemangiomas, they are unresponsive to propranolol. Of all the eight cases reported in Table 2, only two had vascular malformation. Therefore, this is only the third reported case of vascular malformation involving the clitoris and the ninth case overall when considering all the vascular lesions causing clitoromegaly.

Since the hormonal causes of clitoromegaly outnumber other causes, they need to be ruled out first. Radiological assessment in the form of Doppler ultrasound is indispensable for non-hormonal causes as in our case. Once diagnosis is confirmed, both sclerotherapy[8] and surgery have been reported to have successful outcomes. In our case, complete excision of the vascular mass with preservation of the clitoris was possible.

Histopathology confirms the diagnosis and shows the tumour to be composed of large dilated blood-filled vessels lined by flattened endothelium.

Conclusion

Vascular malformation rarely affects the clitoris and may mimic cases of CAH in appearance. Systematic clinical, pathological and radiological examination helps in differentiating hormonal and non-hormonal causes of clitoromegaly and guides cause-specific management.

Consent

Informed consent was taken from the parents of the child for the presentation of her case and clinical pictures. Permission was also taken from the institutional ethics committee.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given their consent for images and other clinical information to be reported in the journal. The parents understand that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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