CASE REPORT

Solitary Vascular Malformation of the Clitoris

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Summary

Isolated involvement of the clitoris by vascular malformation (VM) is very rare. Clinically, the lesion simulates female pseudohermaphroditism. A five-year-old girl presented with clitoromegaly and a clinical diagnosis of solitary VM of the clitoris was made. Magnetic resonance imaging showed characteristic features and confirmed the diagnosis and the extent of the VM. This is the first reported case of isolated involvement of the clitoris by VM to be diagnosed preoperatively.

Key Words: Clitoris, Vascular malformation, Solitary, Clitoromegaly

Introduction

Vascular anomalies are currently classified into hemangiomas and vascular malformations (VM)[1]. Hemangiomas are present at birth, proliferate, and then involute. However, VM are present at birth or develop later but they do not involute. Vascular malformations are errors in morphogenesis that may affect any segment of the vascular tree, including arterial, venous, capillary, and lymphatic vessels, or a combination of the above components. Isolated involvement of the clitoris by VM is rare and in children the resulting clitoromegaly may be misdiagnosed as due to congenital adrenal hyperplasia.

Case Report

A five-year-old girl presented with an enlarged 'phallus' and a history of recurrent bleeding from the lesion in the last few weeks. The lesion was noted in the first month of life and had grown slowly with the growth of the child. The child was born of a non-consanguineous marriage at full-term and had an unremarkable antenatal history. There was no history of any drug ingestion by the mother during pregnancy and there was no family history of similar problems. The postnatal and subsequent growth histories were unremarkable.

Clinical examination revealed a normal looking girl with height and weight appropriate for age. The clitoris was enlarged and looked like a 'penis' of a boy of similar age. The skin over the clitoris was darker than normal with a few dilated veins. There were areas of healed scabs over the skin due to recent episodes of bleeding. There was no bruit over the lesion. The labia majora, labia minora, the vaginal orifice and the vestibule were normal. No other external abnormalities were noted. The cardiovascular system was normal.

A clinical diagnosis of vascular malformation of the clitoris was made. Blood levels of 17-hydroxyl progesterone, estrogens, dihydroepiandrosterone and androstenidione were normal. The karyotype was 46XX. Ultrasonogram showed a vascular mass of tissue in an elongated clitoris. Magnetic resonance imaging (MRI) showed an oval shaped multiseptated lesion in the clitoris, hyperintense on T2 fat saturation image (Fig. 1A) and isointense to muscle in T1 weighted image (Fig.1B). Good enhancement of the lesion was

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seen following gadolinium (Fig.1C) suggesting a well vascularised lesion consistent with the diagnosis of VM. Magnetic resonance angiogram showed normal pelvic vasculature without any major feeding vessel to the clitoral lesion.

Under general anaesthesia, the VM that was confined to the skin and subcutaneous tissue of the clitoris was excised. Both corpora were larger than normal but not involved by the VM. Two small feeding vessels, one on each side, were seen extending from the dorsum of the clitoris into the VM. These were ligated. Reduction clitoroplasty was done preserving the neurovascular supply to the glans of the clitoris. The clitoris was covered with skin over the proximal part of the clitoris. This part of the skin clinically appeared uninvolved by the VM.

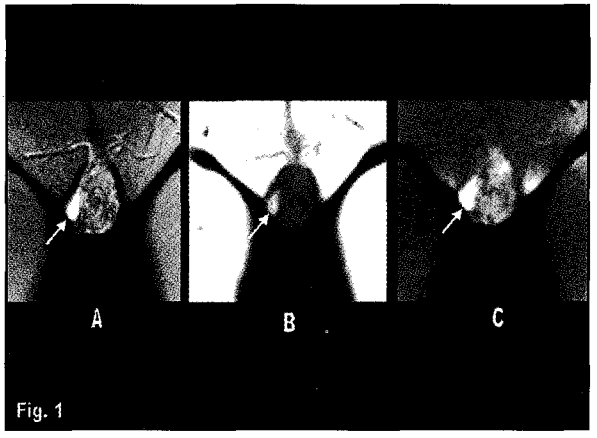

Fig. 1: A - T2 fat saturation coronal MRI. B - T1 image showing lesion isointense to muscle. The area indicated by arrow remains hyperintense, indicating the presence of blood products from a subacute hematoma. C-T1-fat saturation image following gadolinium.

Histology of the excised mass showed multiple endothelial lined, blood filled channels separated by edematous and fibrous connective tissue stroma, confirming the diagnosis of VM. The margin of the retained skin was free.

The patient has been followed up for the past one year and the appearance of the vulva is normal. There has been no evidence of recurrence.

Discussion

Clitoromegaly commonly results from hormonal disorders and intersex states. Hamartomas such as neurofibroma and hemangioma are uncommon causes of clitoral enlargement. Hemangioma of the clitoris usually occurs as part of perineal or pelvic hemangioma. The perineal hemagioma may be associated with anorectal, spinal and genital malformations[2]. Clitoromegaly may be the main presenting feature of genitourinary involvement in neurofibromatosis and is often associated with neurofibromas in the bladder[3].

Solitary involvement of the clitoris by VM closely simulates female pseudohermaphroditism[4]. Prior to this report, a case of isolated involvement of the clitoris by hemangioma has been reported. This patient initially underwent exploration of the inguinal region as a case of intersex[4]. The diagnosis of hemangioma was made at second operation when undue bleeding was encountered and the enlarged tissue was noted to be very soft and spongy. The corpora cavernosa could not be identified and the total mass of enlarged tissue was excised.

In conclusion, a solitary vascular anomaly of the clitoris should be considered in the differential diagnosis of clitoromegaly in children. Correct preoperative diagnosis avoids unnecessary parental anxiety and simplifies management. Correct preoperative diagnosis is possible with awareness of the clinical condition and the specific features seen in MRI.

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