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Infected epidermal cyst of the clitoris in an infant

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ABSTRACT

Clitoral enlargement in the pediatric population is a rare condition, usually related to problems of sexual differentiation, but malignant and benign clitoral lesions have also been described. We report the case of a newborn infant, investigated at birth for an intersex disorder because of clitoromegaly. Hormonal screening was normal and ultrasound (US) did not show a pelvic or abdominal mass. Three weeks later, the lesion was larger, tense and erythematous. An abscess was suspected. A drainage was then performed, and the bacteriological culture revealed the presence of Staphylococci aurei. A magnetic resonance imaging (MRI) performed to exclude a tumor of the soft tissue was normal. A diagnosis of infected epidermal cyst was confirmed by the pathology. Two months later, the external genital aspect was normal and the child asymptomatic.

1. Introduction

A clitoral enlargement can be congenital or acquired. It is diagnosed in children between birth and three years of age when the diameter of the glans is larger than 5mm and the length of the hood superior to 12.6mm [1,2]. It is usually caused by an excess of androgen impregnation during fetal life, infancy or childhood [2]. Tumors of the clitoris such as endodermal sinus tumor, sarcoma, rhabdomyosarcoma, fibroma, leiomyoma, angiokeratoma, hemangioma, neurofibroma and cysts may also occur [2–4]. Cysts are diagnosed in 0.6% of female infants [5]. Numerous other types of pathologies, such as atheromas (from sebaceous glands), dysonogenetic cysts of mesonephric (Müllerian) or paramesonephric (Wolffian) origin and epidermal cysts, can be observed [4,6]. Hormonal screening and radiologic investigations are mandatory to define the lesion as hormonal, non-hormonal, pseudo-clitoromegaly or idiopathic [2]. In most cases, surgery is performed, either to resolve the symptoms or to allow a histopathological diagnosis based on a biopsy. Surgical resection represents a real challenge because of the risk of damaging the neighbouring clitoral components and impairing later sexuality.

2. Case report

A child was born after an uneventful pregnancy with an obvious clitoral enlargement, a 20mm long, solid, rounded mass (Fig. 1). On physical examination, urethral and vaginal meati were present, in a normal position. There was no labial fusion. Gonads were not palpable. There was no sign of virilization. The karyotype was 46XX, and hormonal screening, including 17-OH-progesterone, testosterone, luteinizing hormone (LH), follicle stimulating hormone (FSH), anti-mullerian hormone and alpha-foetoprotein, was also normal. No adrenal mass was revealed by an abdominal and pelvic US, which showed the presence of Müllerian structures, but ovaries could not be seen; the glans of the clitoris measured 13 × 22 mm (Fig. 1). Seven days later, the length of the clitoris was 15 mm. Three weeks after birth, the child was brought to the emergency ward because of the increased size of the clitoris had become larger and was tense and erythematous, resembling an abscess (Fig. 3). Clinical evaluation was otherwise unremarkable, without fever or pain. An emergency surgical drainage was performed; a deep and large cavity extended from the skin along the urethra and vagina. A bacteriological analysis of the purulent discharge showed the presence of a large amount of Staphylococci aurei. The postoperative course was uneventful, with local care (desinfection and soft drain for three days) and oral antibiotics. One month later, the external genital aspect was normal, without clitoromegaly. MRI was finally performed at five months of age to exclude any tumor of the soft tissue (Fig. 2). At seven months, the child was asymptomatic (Fig. 3).

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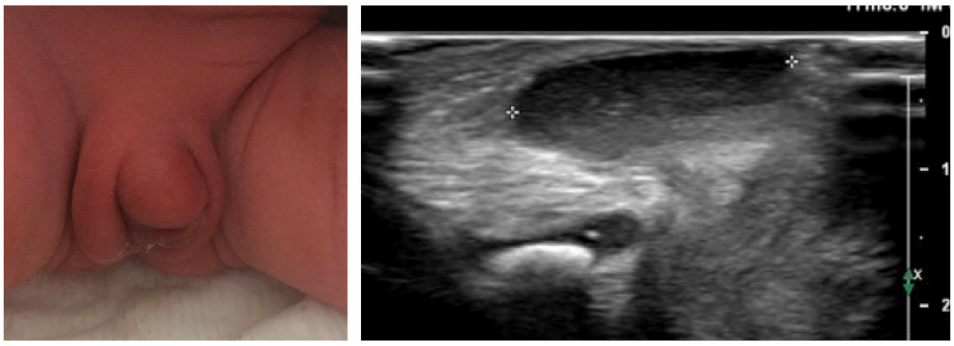

Fig. 1. External genital aspect and US aspect at birth.

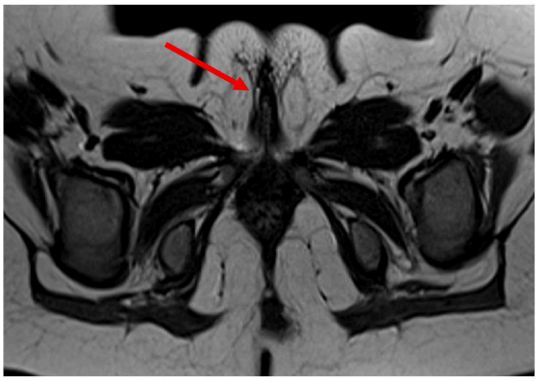

Fig. 2. IRM at 5 months of age.

3. Discussion

Cysts of the external female genitalia are rare and observed in 0.6% of newborn infants [5]. Epidermal cysts occur when the epidermis is displaced into the dermis or subcutis, either dysontogenetically or following a trauma [2,4,6,7]. They are more often described after ritual female genital mutilation [3,4]. A physical examination normally shows a solid, rounded, slowly growing, painless mass [6]. When a cyst is present at birth, it may be misdiagnosed as a problem of sexual differentiation. A hormonal screening should be performed on any patient with clitoromegaly in order to exclude virilization syndromes [2,4,8]: karyotype, 17-OH-progesterone, testosterone, LH, FSH, anti-mullerian hormone and alpha-foetoprotein.

An abdominal and pelvic US should be performed to examine the ovaries and adrenal glands; the clitoral aspect should also be studied to define the nature and length of the cyst [9,10]. MRI is helpful in determining the relationship between the cyst and the clitoris, its potential extension into the pelvis, and its neurovascular component. A cyst is usually described with low signal intensities on T1-weighted images and a high signal intensity on T2-weighted images, but an epidermal cyst is also hyperintense on diffusion-weighted images. MRI is also necessary preoperatively to guide the surgical approach and avoid damages to the glans of the clitoris or the neurovascular bundle [4,10].

The evolution of an epidermal cyst of the clitoris is variable, with either a spontaneous regression [8] or the development of an abscess [7,11]. As our child was very young, we performed a surgical drainage under general anesthesia and an evaluation of the anatomy of the external genitals to make sure that there was no urethral lesion. Broad spectrum antibiotics may be given to try and avoid surgery with its risk of tissue damage in the genital area, as was described in the case of an older girl [11]. In other cases, a histopathology analysis after surgical incision revealed a wall of epidermis with a center filled with keratinaceous material arranged in laminated layers [3,6,7,12–14]. Even if surgery for removal of the tumor is efficacious, precautions should be taken to prevent sensory deficits and alteration of future sexual functioning. Careful dissection and rigorous hemostasis are recommended for the preservation of the glans and the neurovascular bundle [4]. In our case, we performed a late MRI to exclude any tumor, as palpation revealed a persistent mass of fibrous reorganization tissue (scar activity) close to the clitoris. Two months later, as the child was asymptomatic and the mass has regressed, surgical removal cyst was no more indicated and clinical follow-up was decided.

4. Conclusion

An epidermal cyst of the clitoris is a rare condition and may be misdiagnosed as a problem of sexual differentiation. Hormonal screening must be done. US and MRI are useful to narrow down the differential diagnosis. Surgery is mandatory to eliminate the pus and allow a biopsy. When the cyst is removed, the dissection of the tissues must be very carefully performed in order to prevent any future sexual dysfunction.

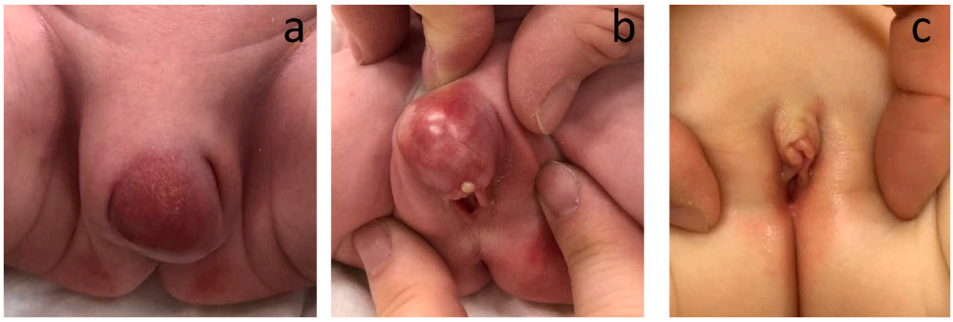

Fig. 3. External genital aspect at 3 weeks of age (a, b) and 7 months of age (c).

Patient consent

Consent to publish the case report was obtained by the parents of the patient. This report does not contain any personal information that could lead to identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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