Case Report

A huge epidermal inclusion cyst of the vulva complicating neonatal female circumcision

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ABSTRACT

Female genital mutilation is the summation of all procedures that involve partial or total removal of the female external genitalia or other injuries to the female genital organs, whether for cultural or other non-therapeutic reasons. Hemorrhage, sepsis and genitourinary damage are the main early causes of morbidity and occasional mortality associated with this practice. Vulva epidermal inclusion cysts, sexual and birth difficulties occur late and can cause medical, psychological and socioeconomic problems. Here, we present the case of a 42-year-old circumcised female, who presented with a huge vulva swelling of 30 years duration that posed some diagnostic challenge because of the presence of other body swellings arising from neurofibromatosis. This was successfully excised and histology showed it was epidermal inclusion cyst, which we believed was a late complication of her neonatal circumcision.

Key words: *Traditional practice, Ritual female circumcision, Female genital mutilation, Neurofibromatosis.*

Female genital mutilation (FGM) was defined by the World Health Organization (WHO) in 1997 as "all procedures that involve partial or total removal of the female external genitalia or other injuries to the female genital organs, whether for cultural or other non-therapeutic reasons.FGM was found in Egyptian mummies as far back as 200 years before Christ (BC). It was practiced in Europe and North America up to the 19th century as a cure for ailments like epilepsy, hysteria, and masturbation [1]. Today, the circumcised females are over 125 million worldwide, primarily in Africa and the Middle East in countries like Nigeria, Ethiopia, Sudan, and Egypt, those in Europe and North America been mostly immigrants [2]. The World Health Organization [3] has classified FGM as Type I, Type II, Type III and Type IV.

Epidermal cysts are formed from the proliferation of epidermal cysts of the skin within a circumscribed space in the dermis and appear as firm, round, mobile cutaneous or subcutaneous nodules of variable sizes. It could be congenital when sequestration of epidermal rests occurs during embryonic life or acquired when there is surgical or traumatic implantation of epidermal tissue or occlusion of a pilosebaceous unit. Here, we report the case of a 42-year-old circumcised female presented with a huge vulva swelling.

CASE REPORT

A 42-year-old female referred to our facility from a peripheral hospital with the complaint of a huge vulva swelling causing dysuria. The practitioner suspected the swelling to be a malignant condition and therefore referred to our hospital. The swelling became noticeable 30 years ago, initially small and symptomless until 10 years ago that it progressively increased in size causing difficulty in micturition with a tendency to push on the swelling and strain each time she desires to urinate. There were other swellings all over the body which started in late childhood, having being told by the parents that she was normal at birth and that she was circumcised by a male traditional healer at one week of age and her wound healed well. She does not have any significant perineal injury since birth and does not have swellings in the bone or any change in bowel habits. Neither the parents nor the sibling had similar skin swellings. In the past, she had visited several hospitals in view of the multiple skin swellings and ugly brown spots but was told nothing could be done to help get rid of them.

She attained menarche at the age of 12 years with a regular menstrual flow to date and she is sexually active. She is from a monogamous family setting and lives with her only younger sister who is married. Both parents are late. A petty trader that deals on fruits and vegetables with the highest level of education being primary education as both parents were poor and she was not doing well in school.

Her physical examination revealed a patient who appears withdrawn, otherwise healthy looking. Vital signs were as follows: Blood pressure 110mmHg systolic and 80mmHg diastolic, pulse rate was 80 beats per minute, respiratory rate was 14breaths per minute

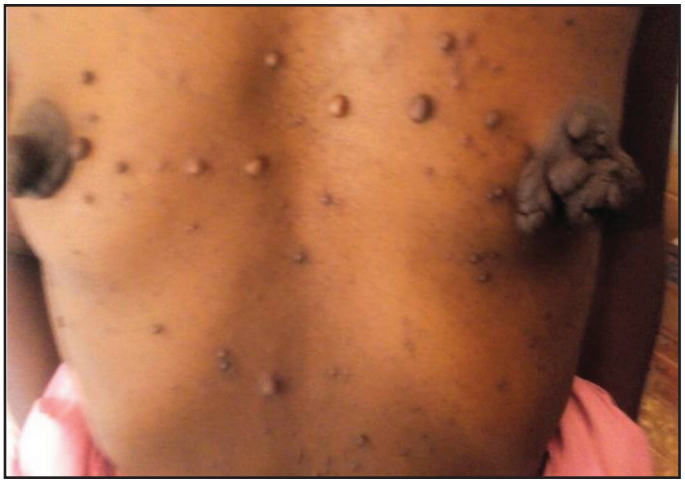

Figure 1: The case showing Café-au-lait spots, fibro-epithelial skin tags and underdeveloped breasts

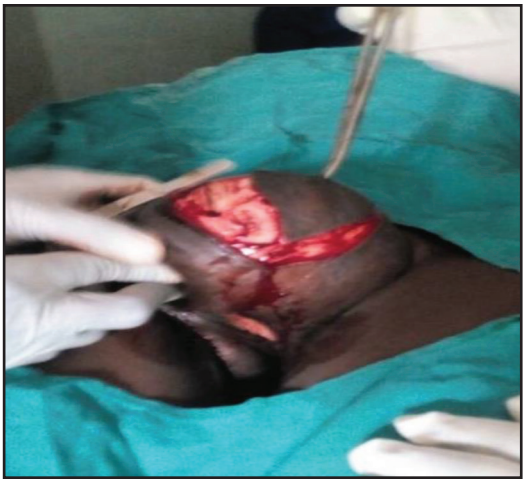

Figure 2: Huge vulva mass, introitus, urethral opening and skin incisions for excision of the mass

and temperature was 36.7°C. Integumentary system examination revealed many Café au lait spots (dark and brown patches), numerous fibro-epithelial skin tags (Fig. 1) and some neurofibromas. Other systems were essentially normal except for the breasts which appeared underdeveloped (Fig. 1) and the perineum which revealed a huge cystic vulva swelling in the region of the clitoris and urethra, non-pigmented and no obvious scar, no differential warmth and measures approximately 20cm in its diameter. It is mobile, non-tender, fluctuant and non-compressible (Fig. 2). Retraction of the swelling showed normal urethral and vaginal openings (Fig. 2). No remarkable finding on digital rectal examination.

Perineal ultrasound scan done confirmed the essential cystic nature of the mass and post-circumcision implantation dermoid cyst was suggested. She was counseled, prepared and had excision biopsy of the mass done in the theatre under sterile condition. A urethral catheter was inserted to prevent injury to the urethra during the surgery, to divert urine and to prevent post-operative narrowing (Fig. 3).

The post-operative period was uneventful and she was discharged after one week. At follow-up visits, she was happy with the cosmetic and functional outcome of the surgery. Histology report revealed an epidermal inclusion cyst.

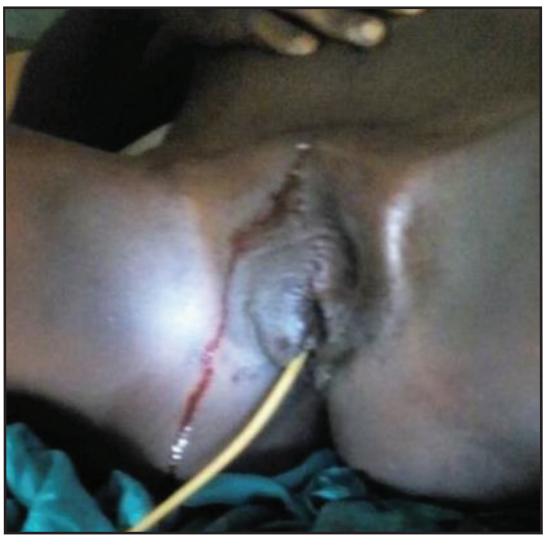

Figure 3: Immediate postoperative appearance of the perineum (urethral catheter in situ)

DISCUSSION

Among the Igbos of South Eastern Nigeria, the traditional practice of female ritual circumcision was prevalent until probably a decade ago when it was accepted as a harmful practice: female genital mutilation (FGM). FGM was classified by the World Health Organization as Type I, Type II, Type III and Type IV [3]. Type I is often referred to as clitoridectomy, this is the partial or total removal of the clitoris (a small, sensitive and erectile part of the female genitals), and in very rare cases, only the prepuce (the fold of skin surrounding the clitoris). Type II is known as excision, this is the partial or total removal of the clitoris and the labia minora (the inner folds of the vulva), with or without excision of the labia majora (the outer folds of skin of the vulva). Type III is referred to as infibulation, which is the narrowing of the vaginal opening through the creation of a covering seal. The seal is formed by cutting and repositioning the labia minora, or labia majora, sometimes through stitching, with or without removal of the clitoris (clitoridectomy) and Type includes all other harmful procedures to the female genitalia for non-medical purposes, e.g. pricking, piercing, incising, scraping, cauterizing the genital area, introduction of corrosive substances or herbs into the vagina for non-medical purposes. Types I and II are most represented in about 80% of FGM while type III, though most severe, is found in minority of cases [4].

Early complications following FGM include hemorrhage, urinary retention, severe pain, shock, infections including tetanus which can lead to mortality. Late complications may include vulva cyst, urogenital infections, pelvic inflammatory disease, hypertrophic scars, dyspareunia, infertility and obstetric problems (like delayed or obstructed labor, hemorrhage). These can cause physiological, psychological and socioeconomic problems [4, 5]

The traditional practice of circumcision in many places in Africa was rooted in culture and tradition and justified in the belief that it attenuates sexual desires in females, thereby ensuring chastity and fidelity [6,7]. This practice has been condemned

by the WHO [8] and the practice outlawed in many countries including Nigeria [9].

Epidermal inclusion cyst following female circumcision is rare considering the prevalence of circumcised females in Nigeria [41%] [10] and therefore can cause misdiagnosis especially when it becomes very large or is associated with other cutaneous swellings. The case presented was large and occurred in a patient with Von Recklinghausen's disease (neurofibromatosis type 1). The referring physician wanted to be sure that this was not a sarcomatous change in a neurofibroma, which is known to occur in 5% of patients with this disease [11].

With the benefit of experience, the history of ritual circumcision in infancy, the site of the mass near the clitoral area, its cystic nature which was confirmed by ultrasound, we were confident we were most likely dealing with a benign swelling before the surgery, probably an epidermal inclusion cyst. Many reports in the medical literature have discussed similar swellings following traditional circumcision in both females [12, 13] and males [14, 15]. It has also been reported following blunt trauma in the vulva [16,]

The pathogenesis of inclusion cysts along the edges of any surgical procedure includes smegma accumulating in the incision or from epidermis rolling in at the time of the procedure [17]. The long history before presentation in our index patient may be occasioned by her socioeconomic background of poverty and poor education. Treatment aims to excise the cyst and reconstruct a cosmetically acceptable vulva with normal urethral and vaginal orifices [18]. A complication which is common in such cyst in other parts of the body is an infection, which is usually treated initially by incision and drainage, antibiotics cover and interval excision of the mass. A malignant change to squamous cell carcinoma in an epidermal cyst has been described [19] and radical surgery will be required.

Usually, these cysts grow very slowly and are usually less than 5cm in diameter when diagnosed [20]. Large cysts are cosmetically unacceptable and may interfere with micturition and sexual functions.

CONCLUSION

The diagnosis of epidermal inclusion cyst should always be considered in any cutaneous swelling with a scar; swelling around the genital in circumcised individuals should not be an exception. Clinical diagnosis should not be too difficult with a history of trauma (FGM), a cystic lesion confirmed by perineal ultrasound and the benefit of previous experience. Despite the long history, the huge size of her vulva swelling and other medical conditions of our index patient, we are happy to say that she was satisfied with the final cosmetic and functional outcome of the treatment.

Gratitude to Almighty God who gave us the privilege to embark and complete this project, also to the patient who gave her consent to publish this report.

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