

The largest epidermal cyst with vitiligo lesions following female genital mutilation: a case report and literature review

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Abstract

Epidermoid vulvar cystic lesions are proliferations of epidermal cells that can occur as a complication of female genital mutilation (FGM), which is still a common practice in many cultures, especially in Africa. A 36-year-old Sudanese woman presented with an enlarged clitoral mass that had first appeared 2 years earlier. Her medical history showed that she had undergone FGM when she was 3 years old. A perineal examination revealed a mobile, nontender, rounded cystic swelling with vitiligo lesions. After the cyst was excised, it revealed a 13 × 11 × 11 cm unilocular round mass. An epidermoid cyst was reported following microscopy. Follow-up 6 months later revealed a good result with no recurrence. To date, this is the largest epidermoid cyst following FGM and the first one with vitiligo lesions reported in the literature.

Keywords: epidermal cyst, vitiligo, female genital mutilation

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Introduction

Cystic lesions such as epidermal vulvar cysts are extremely rare without a prior surgical operation following trivial trauma, infibulation, circumcision, or genital piercing (1, 2). The majority of cases occur as a complication of female genital mutilation (FGM), which is still a common procedure in many cultures—especially in Africa—on neonates (1–14).

FGM has been practiced for centuries in more than 30 countries and is most prevalent in African countries such as Nigeria, Ethiopia, Sudan, and Egypt, and in some areas of the Middle East (14). FGM is classified by the World Health Organization (WHO), and type III (removal of the clitoris and labia minora and narrowing of the vaginal orifice) is the most common procedure performed in some countries (15–17). Various complications are observed after circumcisions, such as epidermal cysts (18).

This case report is of a 36-year-old woman with a history of type III FGM and a giant clitoral epidermal cyst accompanied by vitiligo lesions. To date, this is the largest epidermoid cyst following FGM and the first one with vitiligo lesions reported in the literature.

Case report

A 36-year-old Sudanese woman presented with an enlarged and painful clitoral mass with vitiligo lesions that was causing sexual difficulties and psychological distress. Her husband suspected that she had been bewitched into having a single scrotum (Fig. 1). The enlargement had been present for 2 years and had grown rapidly within the previous 6 months. The mass had started growing larger and causing a dragging sensation in the vulva and significant discomfort, especially during intercourse, but the patient had not experienced any inflammation or infection of the mass. She did not have a history of cystic lesions following trauma or surgery at any other sites. The patient had had vitiligo lesions in the same location as the cyst for 1 year. Over the course of the preceding year, the vitiligo lesions had increased in diameter in the

same proportion as the cystic mass. The initial vitiligo lesion was on her umbilical scar following gastrointestinal surgery 3 years earlier. She did not have any additional vitiligo lesions except on the mutilation scar. The patient had no history of autoimmune disease. Her family history was negative for vitiligo.

The patient had had regular cyclic menses since she was 13 years old. She had no history of birth, miscarriages, or genital piercing. She did not have a history of urogenital infections. Her medical history included type III FGM at the age of three, performed by a village midwife. A perineal examination revealed a 13 × 13 cm mobile, nontender, rounded cystic swelling in the clitoral area, which was covered with normal skin other than the depigmented skin touching the medial thighs and vestibulum (Fig. 1). The cyst was neither fixed to the symphysis pubis nor involved



Figure 1 | Clinical and postoperative picture of the giant epidermoid cyst with vitiligo lesions.

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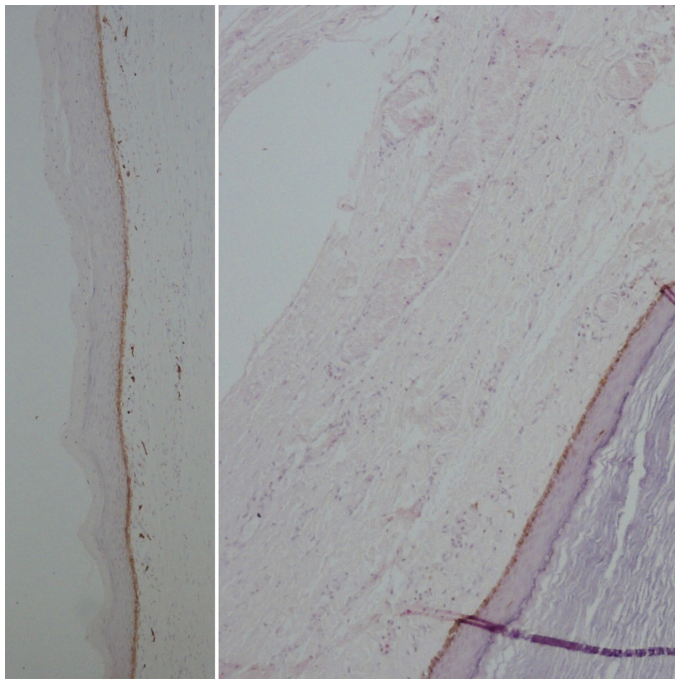


Figure 2 | Histopathological appearance of the epidermoid cyst lined by an epidermis-like epithelium including a granular cell layer and filled with laminated keratin.

Discussion

Various types of cysts can occur in the clitoral region, including atheroma, dermoid cysts, dysontogenetic cysts, and epidermoid cysts (14). Such cysts can cause clitoromegaly and can be described as ambiguous genitalia when present at birth (19). Many of these cysts resolve spontaneously (20), but in this case there was a late onset of ambiguous genitalia that grew within months.

Following FGM, epidermoid cysts are mainly seen in the clitoris, labia majora, and labia minora, at various rates according to the type of circumcision (1, 8, 21, 22). According to the WHO, two million new FGMs are performed annually in the world (16, 23). Circumcisions are usually performed on neonates under traditional conditions, often by older women that are midwives (24, 25). The patient described in this report had a female circumcision when she was 3 years old.

FGM is classified by the WHO into four types (16, 26, 27). These are Type I: removal of the prepuce with or without removal of all or part of the clitoris; Type II: removal of the prepuce, clitoris, and part of the labia; and Type III: removal of all or part of the external genitalia along with stitching or narrowing of the vaginal opening (27). This kind of mutilation, Type III, also called infibulation (5), is the most extreme type. Incisions are made in the labia majora to create raw surfaces that are then either stitched together or kept in close contact until they seal and form a cover for the urethral meatus. A very small orifice is left for the passage. In many regions, this is the most common procedure performed; for example, 98% of FGMs are Type III in Djibouti and Somalia (15–17). Various complications are seen after circumcision, such as bleeding, infection, shock, menstrual problems, difficulties with urination, common urinary tract infections, urinary retention, tetanus (which can lead to mortality), inguinal pain, difficulties with sexual intercourse, a genital circumcision scar—especially at the vulvar region—and a cystic or solid mass of variable size, as found in this patient (18).

Epidermoid cysts following FGM are not rare. A review of the

literature is summarized in Table 1. According to the literature, the time from circumcision to the development of the cyst is variable, but a delay of more than 30 years—as in the present case—is very infrequent (10, 12, 13). In 2010 Asante et al. reported a 37-year-old female from Guinea with a large clitoral epidermal inclusion cyst of the clitoris 30 years after FGM with a history of 6 months (10). In 2015 Birge et al. reported a female from Nyala, Sudan with a vulvar epidermoid cyst after FGM. They reported an interval of 35 years between FGM and the surgical intervention (12). Finally, in 2016 Victoria Martinez et al. reported a 37-year-old female patient from Nigeria that developed a cyst more than 30 years after FGM (13). Similar to these previous reports, the patient in the case reported here had a cyst 31 years after FGM.

The largest epidermoid mass that developed in the labium major was reported by Yang et al. and was about 12 cm in size with 10 years of clinical history (28). From this perspective, a mass of 13 cm accompanied by vitiligo lesions with a delay of 31 years in presentation is the largest epidermoid mass with peculiar characteristics reported in the literature.

The case presented in this paper is different from previous reports in that it involves vitiligo lesions. Vitiligo is an acquired, chronic, depigmenting disorder of the skin with an estimated prevalence of 0.5% of the general population. It is characterized by the development of white macules on affected areas. Multiple mechanisms are involved in vitiligo; namely, genetic predisposition, environmental triggers, metabolic abnormalities, impaired renewal, and altered inflammatory and immune responses (29). In this patient, vitiligo lesions likely occurred due to the Koebner phenomenon. The Koebner phenomenon refers to the development of isomorphic lesions in the traumatized, uninvolved skin of patients that have skin diseases, including psoriasis, vitiligo, and lichen planus. These injuries can be non-penetrating blunt trauma—such as stretching, friction, compression, and vibration—similar to this patient's genital trauma (30). In addition, the patient had a history of an old and healed vitiligo lesion on her umbilical scar from gastrointestinal surgery; this further supports the development of vitiligo lesions as a consequence of the Koebner phenomenon.

Table 1 | Review of reports of patients with epidermoid vulvar cysts after female genital mutilation.

Year	Author (Reference no.)	Country of origin	Patients (n)
1992	Dirie and Lindmark (5)	Somalia	290
1995	Hanly and Ojeda (6)	Saudi Arabia	10
1999	Baaij et al. (7)	Somalia	1
1999	Abudaia et al. (19)	Saudi Arabia	1
1999	Adekunle et al. (8)	Nigeria	39
2001	Rouzi et al. (4)	Saudi Arabia	21
2004	Yoong et al. (9)	Somalia	1
2010	Asante et al. (10)	Guinea	1
2010	Rouzi (11)	Saudi Arabia	15
2010	Osifo (2)	Nigeria	37
2015	Birge et al. (12)	Sudan	1
2016	Victoria-Martinez et al. (13)	Nigeria	1
2018	Kibar Ozturk, present case	Sudan	1

Conclusion

This report describes a 36-year-old woman with a sizeable (13 × 11 × 11 cm) clitoral epidermal cyst accompanied by vitiligo lesions secondary to FGM. FGM is still common in Africa and among immigrants from Africa, especially in culturally and traditionally closed societies. More investigations are warranted to elaborate on the short- and long-term complications of FGM.

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