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Case Report

Vulvar Schistosomiasis: An Important but Uncommon Diagnosis: "A Near Miss"

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ABSTRACT

Background: Vulvar schistosomias, part of the spectrum of female genital schistosomiasis (FGS), is an important tropical disease with far-reaching negative impact on female reproductive health. However, this disease is rarely diagnosed and reported in clinical even in endemic tropical Africa like Nigeria due to poor awareness and low index of suspicion among the physicians in the tropics. This could result in avoidable morbidities. **Case report**: We report a successfully managed case of vulvar schistomsomiasis that was long misdiagnosed and mismanaged by unsuspecting physicians, and was nearly misdiagnosed by us. **Conclusion**: Female genital schistosomiasis, an important tropical disease, is rarely diagnosed in clinical setting even in the endemic regions resulting in avoidable morbidities. This case report calls attention of the physicians especially in the tropics to think outside the usual in their approach to diagnosis and management of female genital pathologies. The need for biopsy of any suspicious lesion was also underscored.

INTRODUCTION

Schistosomiasis is a water borne parasitic disease caused by a trematode worm of the genius Schistosoma. Also referred to as bilharziasis or snail fever, the disease was first described in Egypt by Theodor Bilharz in 1851.[1]

Schistosomiasis is said to be the third most devastating tropical disease in the world, being a major source of morbidity and mortality for developing countries.[1] The major schistosome species include Schistosoma haematobium, which is the most widespread in Sub-Saharan Africa and responsible for urinary schistosomiasis, while Schistosoma mansoni, S. intercalatum, S. japonicum and S. mekongi are all responsible for intestinal schistosomiasis.[2]

About 230 million people in 74 countries (90% of whom in Africa) are infected worldwide and at least 600 million are at risk of infection.[3,5] An estimated 120 million suffer severe consequences of the infection with an estimated annual mortality rate of about 20,000 worldwide.[3] An estimated 30 million Nigerians need to be treated annually for the disease.[4]

The geographic distribution and etiology of schistosomiasis reflect the unique life cycle of Schistosoma species. Schistosomes infect susceptible freshwater snails in endemic areas, usually with specific species of schistosomes infecting specific species of snails. The infected snails release cercariae 4-6 weeks after infection. They can survive in fresh water up to 72 hours, during which time they must attach to human skin or to that of another susceptible host mammal or die.[5]

Schistosomiasis can affect the skin by three mechanisms. In schistosome dermatitis (swimmer's itch), cercariae, usually of avian species, penetrate the skin, causing a localised allergic reaction. Itchy papules and urticaria occur within one to two hours of swimming in fresh or salt water.[6] Secondly, mature worms may be associated with erythematous itchy macules at the time of release of a large number of eggs due probably to a systemic hypersensitivity reaction to schistosome antigens.[6] Thirdly and most commonly, skin diseases usually around the genitalia results from a chronic inflammatory reaction to the deposition of ova in the skin.[7]

Female genital schistosomiasis (FGS), including vulvar schistosomiasis, is a frequent complication in women with urinary or systemic schistosomiasis, particularly in geographic areas where the disease is endemic.[3, 8] Nigeria is one of the schistosomiasis endemic zones in tropical Africa but there are not many reported cases of FGS in clinical setting.[14] This is despite the far-reaching negative impact of FGS on the female reproductive health. The authors believe lack of awareness and low index of suspicion could account for low diagnosis and reportage of this important but neglected tropical disease. This underscores the need for the case report. It calls attention of the physicians to think outside the usual in their approach to diagnosis and management of female genital pathologies.

We report a successfully managed case of vulvar schistomsomiasis in a young woman that was long misdiagnosed and mismanaged, and was nearly missed by us.

Patient consent was obtained and approval given by hospital ethical committee to report the case.

CASE SUMMARY

Patient was a 23-year-old graduate who presented with 6 months history of vulvar itching and swelling. Itching was intermittent but intense, causing her significant discomfort and occasional embarrassment. There was no known aggravating factor but temporarily relieved by scratching. Scratching could be vicious, a times leading to bruising. Vulva swelling was noticed about the same period, prior to presentation. Swelling was insidious in onset, located around the labia minora. It felt rubbery, occasionally rough but was not painful. There was no associated abnormal vaginal discharge or odor. No urinary symptoms was reported by patient. Her developmental milestone and puberty were normal. Patient, at presentation, was at her ovulatory cycle.

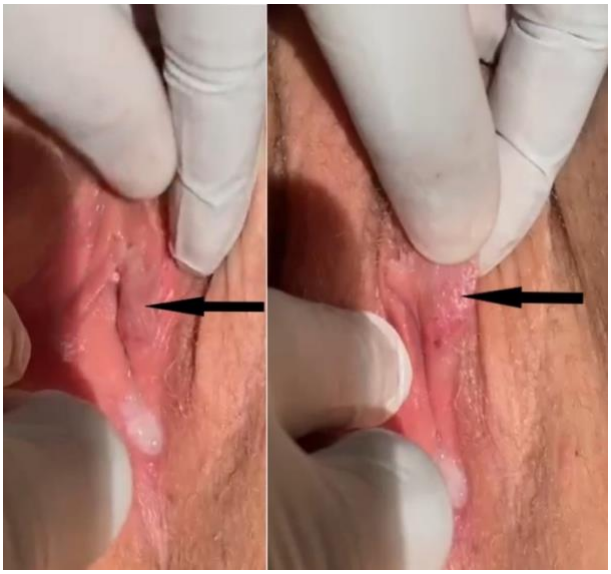


Figure 1. Clinical photograph of the warty papillomatous vulvar lesion

She was single, and had not engaged in penetrative vaginal intercourse, though she admitted to occasional oral sex and superficial genital contact with her male partner. Patient social class could be placed as above average. She recently

graduated from a tertiary institution where she lived with 2 other students in a self-contained one room apartment in the northern part of Nigeria. No history of travel to a high endemic schistosomiasis zone.

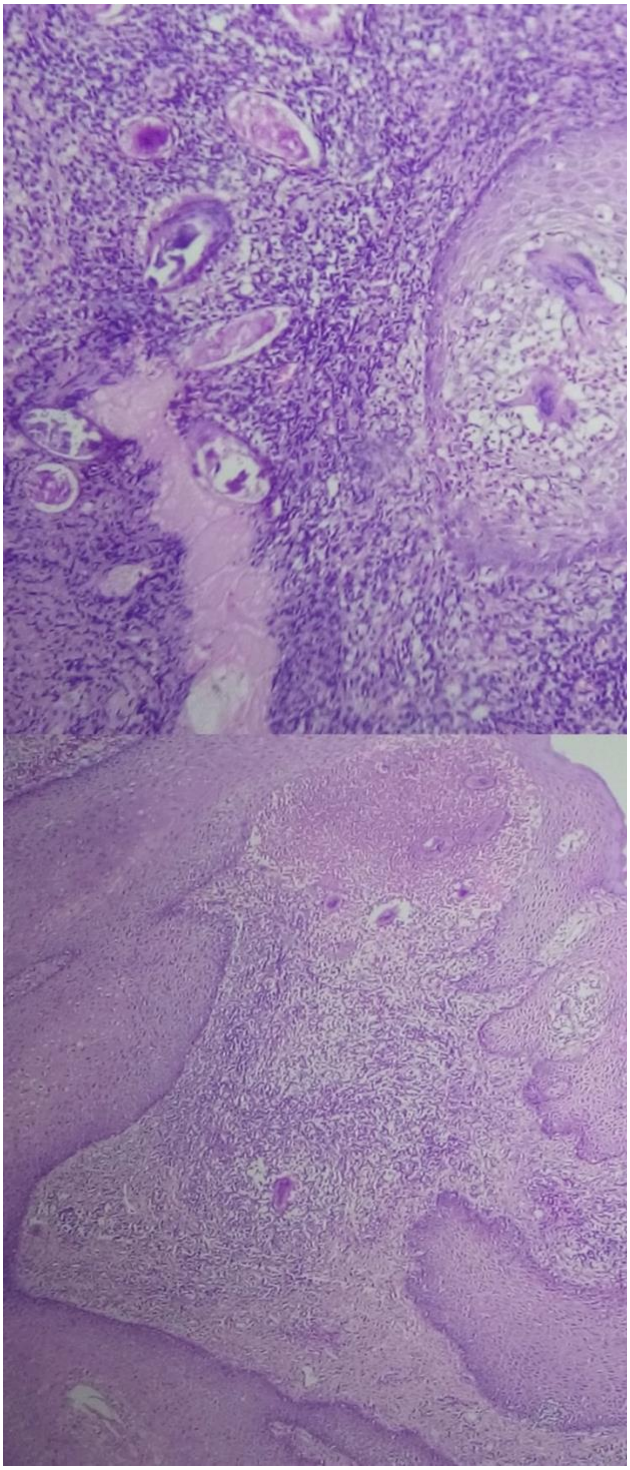


Figure 2. Photomicrograph of the histology of the vulvar lesion

She had indulged in self-medication with a lot of topical creams and vaginal inserts to no avail. She also patronized laboratories for investigations involving blood, urine and vaginal fluid samples. Patient had seen a few gynaecologists who made impressions ranging from "pruritus vulvae" "genital wart" to "psychologically related causes". Medications, both oral and topical including counselling were offered to patient with no good result. She came to a reference hospital in southern Nigeria for test of medical fitness, and was referred from the general outpatient department to the gynecologist on account of her symptoms, and thus she presented to the gynaecology clinic.

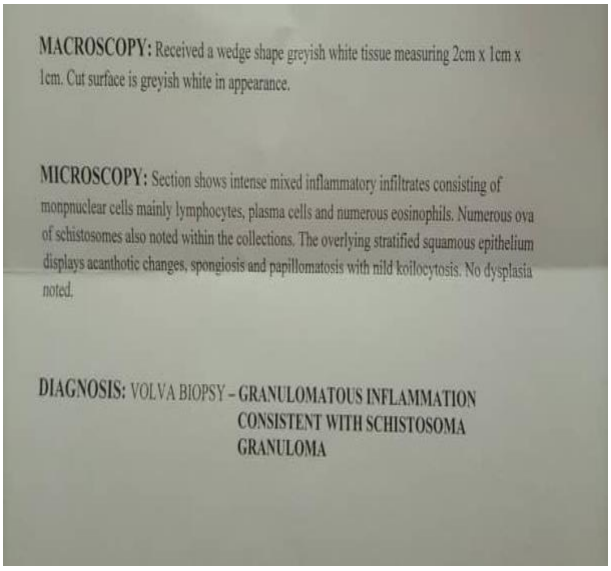
On examination at presentation, she was generally a healthy looking but worried young woman. All vital signs were within normal limits. Secondary sexual characteristics were normal for age.

Examination of the external genitalia revealed a fleshy, partly warty and papillomatous lesion on the upper lip of the left labia minora extending to the left lower base of the clitoris. Minor bruises were noted on the lesion (possibly due to scratching). The contralateral labia and other parts of the external genitalia were essentially normal. The hymen was grossly intact. Mucoid (probably ovulatory) discharge was noted on the vaginal introitus.

An impression of vulva wart was made, and plan was to place patient on topical podophylin cream. However, based on history and suspicious findings, decision was taken for biopsy. This was done under local anesthesia. Histological report revealed "intense mixed inflammatory infiltrates consisting of mononuclear cells mainly lymphocytes, plasma cells and numerous eosinophils. Numerous ova of schistosomes also noted within collections. The overlying stratified squamous epithelium displays acanthotic changes and papillomatosis with mild koilocytosis. No dysplasia noted. Diagnosis was "granulomatous inflammation consistent with schistosoma granuloma." Urine analysis and microscopy were essentially normal

The patient was counseled on findings. She was started on oral praziquantel 40mg/kg in 3 divided doses. Patient reported very significant relief of symptoms 2 weeks on follow up. The vulvar lesion also had markedly resolved. She was

given a repeat dose and a 4-week appointment but she didn't show up.


MACROSCOPY: Received a wedge shape greyish white tissue measuring 2cm x 1cm x 1cm. Cut surface is greyish white in appearance.

MICROSCOPY: Section shows intense mixed inflammatory infiltrates consisting of monpnuclear cells mainly lymphocytes, plasma cells and numerous eosinophils. Numerous ova of schistosomes also noted within the collections. The overlying stratified squamous epithelium displays acanthotic changes, spongiosis and papillomatosis with mild koilocytosis. No dysplasia noted.

DIAGNOSIS: VOLVA BIOPSY – GRANULOMATOUS INFLAMMATION CONSISTENT WITH SCHISTOSOMA GRANULOMA

Figure 3. Histology Report

DISCUSSION

Vulvar schistosomiasis is not a common diagnosis even in endemic regions.[1, 4] It is part of the spectrum of female genital schistosomiasis (FGS) defined as the presence of ova and/or a characteristic pathology in female reproductive organs.[9] FGS remains highly prevalent and under-diagnosed due to a low index of suspicion among health-care professionals.[10, 11]

Female genital schistomiasis is a manifestation mainly of the species S. haematobium infection. Several studies have strongly associated FGS to pathologies of the female reproductive system including subfertility, infertility, and ectopic pregnancies, pregnancy complications (abortion, still-birth and preterm delivery and low birth weight).[1] Tubo-ovarian adnexae masses, both benign and malignant associated with schistosomiasis have been described in several histopathological series[3, 12]

The uterine cervix is the female genital organ most frequently affected by schistosomiasis.[3, 11] Several clinical signs and symptoms have been significantly associated with cervical schistosomiasis such as low abdominal pain, irregular menstruation, cervical contact bleeding, abnormal vaginal discharge, dysparunia, cervical polyps, chronic cervicitis.[12]

Vulvo-vaginal pathology due to schistosomiasis has been reported for over a century. Indeed, the first FGS reported in 1899 was a case of a warty prominent mass in the vagina of an Egyptian woman.[3] Vulvar schistosomiasis lesions can be easily confounded with condyloma acuminata (genital warts due to Human Papilloma Virus (HPV)), and polypoid/papillomatous tumours, in the vagina and vulva, can be considered to be pathognomonic for FGS in the absence of HPV infection or syphilis.[3, 12]

Our index patient presented with vulvar lesion grossly consistent with vulvar wart the possible reason for the misdiagnosis even by unsuspecting gynaecologists. We nearly missed the diagnosis but for more indebt history (which was not strongly suggestive of HPV infection), thinking outside the usual, and timely decision to biopsy the lesion for histology. It could be possible that the primary infection, FGS, being a risk factor for HPV infection[12], may have possibly predisposed the patient to genital wart due to HPV infection. However, low risk of HPV infection by history, non-response to typical treatment to genital wart, rapid response to praziquantel treatment, and the histology report resolves the primary diagnosis in favor of FGS.

The clinical appearence of vulval schistosomisais includes progressive and relapsing swelling, painful or painless ulceration, papules, nodules, pruritus, a hypertrophic clitoris with an eroded granular surface and papillomatous lesions forming masses resembling condylomata.[10,11] Some of these clinical features were present in our index patient.

For over 2 decades, strong associations between HIV and HPV infections and FGS have been made in several studies.[12,13 15] FGS increases susceptibility to HIV infection through disruption of the vaginal and cervical epithelium caused by erosion or inflammation. As in trauma or ulcerative sexually transmitted diseases, neovascularisation and disruptions in the integrity of the epithelial barrier are associated with an increased risk for HIV infection. The friable epithelium and bleeding during coitus in women with FGS facilitates access to deeper genital cell layers by HIV in semen. Schistosomiasis also can alter the immune responses to HIV. The immunoregulatory responses associated with helminth infection downregulate the T-helper-1-type immune response associated with

control of viral infections. HIV replicates more readily in the T-helper-2-type cells associated with helminth infections.[15] Controlling schistosomiasis may reduce risk of HIV among women and contribute to controlling the HIV epidemic in sub-Saharan Africa.

The relationship between FGS (especially cervical schistosomiasis) and cancer of the cervix is not far-fetched. A recent longitudinal study showed that the development of high-grade squamous intraepithelial neoplasia was significantly associated with FGS of a minimum of 5 years duration.[12]

The association of FGS and numerous obstetrics and gynaecological pathologies has been highlighted in medical case reports for over a century. The vast majority are from women living in schistosomiasis endemic countries or from individual cases of tourists who acquired FGS in sub-Saharan countries and were diagnosed in the well-equipped health care facilities of western countries.[12]

Few cases of FGS in clinical setting have been diagnosed and reported from schistosomiasis endemic regions including Nigeria. This could be attributed to low index of suspicion among physicians, lack of reliable diagnostic facilities and trained personnel, lack of integrated approach to diagnosis and treatment. Hence, a lot of such cases may have been neglected, misdiagnosed and mismanaged as other common genital pathologies that physicians are used to. This could result to significant but avoidable morbidities and mortalities.

CONCLUSION

Female genital schistosomiasis is an important but neglected tropical disease. Clinical diagnosis and reportage are few even in an endemic zone like Nigeria possibly due to poor awareness and low index of suspicion. The authors believe this case is one of numerous cases of FGS that may have been missed, misdiagnosed and mismanaged, and hence calls attention for high index of suspicion. It also emphasizes the need to biopsy every suspicious lesion without which this case would have been missed.

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