Case Study

A case of vulval schistosomiasis

O'Mahony D, MBBCh, DCH, DTM&H, DPH, DIP MID COG (SA), FRCGP
Family Practitioner, Bridge Street, Port St Johns
Banach L, MD, PhD, MIAC
Associate Professor, Department of Pathology, Faculty of Medicine & Health Sciences, Walter Sisulu University, Mthatha
La Grange LL, MBBCh, MMed (Derm)
Specialist Dermatologist, East London Hospital Complex, East London

Correspondence to: Dr D O'Mahony, e-mail: omahonyd@softdux.co.za

Abstract

While schistosomiasis is endemic in South Africa, cutaneous manifestations are relatively uncommon. This case report describes the clinical findings in a patient with vulval schistosomiasis. Schistosomiasis should be considered in the differential diagnosis of genital lesions in residents in endemic areas or visitors to these areas.

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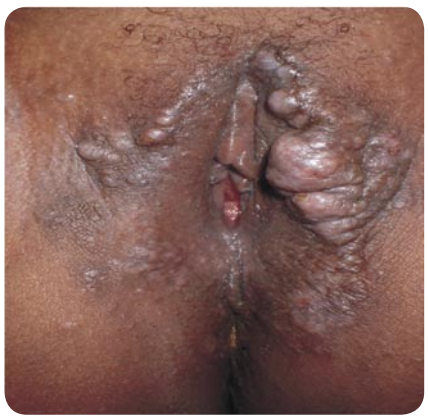
Case Study

An 11-year-old girl, resident at Qandu, Port St Johns, presented to her general practitioner (GP) in June 2006 with an itchy genital rash of one week's duration. There was no history of sexual abuse and she was in good general health. On examination, there were papules and nodules in the labia majora (Figure 1). The GP formulated a differential diagnosis of lichen planus, molluscum contagiosum or condylomata accuminata. As the features were not typical, he obtained a teledermatology consult via the iPath server based at Walter Sisulu University.[1] The dermatologist reported that there was prominent lichenification and hyperpigmentation, which suggested a longer history. The lesions on the right looked more eczematous, with the lesions on the left that may be lichen planus like, but not typical. The nodules did not fit with either, or with molluscum contagiosum. She suggested a biopsy, which revealed ova of *S. haematobium* in the epidermis and underlying tissue, surrounded by a heavy chronic inflammatory cell infiltrate.

Figure 1: Appearance of vulva and perineum



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.[2] Itchy papules and urticaria occur within one to two hours of swimming in fresh or salt water. Secondly, mature worms may be associated with erythematous itchy macules at the time of release of a large number of eggs. This is probably a systemic hypersensitivity reaction to schistosome antigens from the main schistosomes of man (*S. mansoni*, *S. japonicum* and *S. haematobium*). A similar mechanism causes erythema, macules and pruriginous lesions in Katayama fever. Thirdly and most commonly, skin disease results from a chronic inflammatory reaction to the deposition of ova in the skin. Most skin lesions occur around the genitalia.[2]

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.[2,3] Fistulae ('watering can perineum') may occur. Diagnosis can be made by biopsy or from the wet examination of a tissue scraping. The biopsy can be examined by the quantitative compressed biopsy technique, where the material is compressed between two glass slides, or by routine histology.[3]

Vulval schistosomiasis is not a common diagnosis in the endemic regions of South Africa. In the period 1986 to 2000, only 20 cases were reported to the Department of Anatomical Pathology at Walter Sisulu University (WSU), Mthatha, where most histology specimens from the Transkei region are processed (AJ Stepien, personal communication 12/03/2007). From 1994 to 1998, 27 cases of vulvar schistosomiasis were diagnosed on histology at the Department of Anatomical Pathology at the Nelson R Mandela School of Medicine, Durban.[4] At WSU, the diagnosis of vulval schistosomiasis was made on histology and was not suspected by the referring physician. No details were provided in the Durban study.

Vulval schistosomiasis is part of the spectrum of female genital schistosomiasis (FGS). It is defined as the presence of ova and/or a characteristic pathology in reproductive organs.[3] This is a common manifestation of infection with *S. haematobium*. *S. mansoni* may also cause FGS in endemic areas. Possible consequences include facilitation of HIV and other sexually transmitted diseases as a result of thinning, erosion and ulceration of the epithelium of the cervix. FGS may impact adversely

on the reproductive health of women, causing infertility, ectopic pregnancies and pregnancy complications (abortion, still-birth and premature delivery). In population-based studies in countries with areas of endemic schistosomiasis, the point prevalence's of FGS of the lower genital tract have ranged from 33 to 75%.[3] There is a need to raise awareness of this disease entity and to consider it as a differential diagnosis in routine gynaecological practice.

Schistosomiasis is endemic in the Port St Johns district. A survey of the prevalence and intensity of *S .haematobium* urinary infection in schoolchildren and adults in the district, undertaken in the years 1987 to 1989, found an overall prevalence of 42% (range 10% to 90%).[5] Infection intensities are expressed as eggs per centilitre (EPC, 1 centilitre = 10 ml). While the authors of this survey defined a high-intensity infection as ≥ 200 EPC of urine, the World Health Organization (WHO) now defines heavy-intensity infections as ≥ 50 EPC.[6] Using the WHO criteria, the majority of schoolchildren with schistosomiasis had heavy-intensity infections. The WHO recommends that targeted treatment with praziquantel be given in all endemic areas, with the re-treatment interval being determined by the baseline prevalence rate. Since schoolchildren usually have the highest prevalence and intensity of infection, the WHO recommends that they be targeted in control programmes. The emphasis now is on reducing and reversing morbidity, especially in those most heavily infected. More permanent control can be achieved only by improvements in sanitation, access to clean water, snail control and health education. Even without these additional measures, however, drug treatment brings immediate benefit and has a long-lasting effect on morbidity, preventing irreversible sequelae in adulthood.[6]

There is no programme in the Eastern Cape to treat schoolchildren in the affected areas. Praziquantel treatment has been shown to reduce the burden of urinary schistosomiasis in rural KwaZulu-Natal, where the Department of Health, in cooperation with the Department of Education, established a pilot programme in 1998 that aimed to regularly treat primary school children for schistosome and intestinal helminth infections.[7] However, the programme was discontinued in 2000 (C Appleton, personal communication 12/01/2007). As a member of the World Health Assembly, South Africa supports resolution number WHA54.19 of 2001, which sets the goal of attaining a minimum target of regular administration of chemotherapy against schistosomiasis (and soil-transmitted helminth infections) to at least 75% and up to 100% of all school-age children at risk of morbidity by 2010. Plans for such a programme are currently being finalised by the Directorate of Child and Youth Health at the Department of Health (L Bamford, personal communication 14/03/2007). This case is a reminder that schistosomiasis remains a public health problem that needs continued control.

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