Late Cutaneous Schistosomiasis Representing an Isolated Skin Manifestation of Schistosoma mansoni Infection

G. Kick  M. Schaller  H.C. Korting

Department of Dermatology, Ludwig Maximilian University, Munich, Germany

Abstract
Ectopic late cutaneous schistosomiasis is usually preceded or accompanied by visceral schistosomiasis infection. Our patient presented the very rare case of late cutaneous schistosomiasis as an isolated skin manifestation. Perigenital lesions occurred 1 year after contact with infested water. Identification of the few eggs remaining in the late lesion among the dense cellular infiltrate was difficult. Electron-microscopic studies clearly demonstrated the characteristic eggshell ultrastructure.

Case Report
During a 1-year travel through Africa, a 35-year-old man enjoyed swimming in Lake Malawi and the Niger River. One year later, he developed a slowly growing and initially slightly itching tumour in his right groin. There was no skin eruption and no history of haematuria or cystitis, bloody diarrhoea or dysentery during his stay in Africa. Physical examination showed a broadly based reddish tumour in the right groin, partly with a glassy surface (fig. 1). Regarding the clinical findings, pyogranulomatous inflammation and fibrosis. Intermittent abdominal pain, diarrhoea, hepatosplenomegaly and portal and pulmonary hypertension are consecutive symptoms. Other sites of oviposition, as presented by cutaneous lesions, are rare. Our case report demonstrates the clinical, histological and ultrastructural findings, linked to late cutaneous schistosomiasis.

Key Words
Cutaneous schistosomiasis - Schistosoma mansoni - Bilharziasis cutanea tarda

Introduction

The denomination schistosomiasis indicates a group of diseases caused by trematodes of the genus Schistosoma [1]. In particular, the life cycle of the species Schistosoma mansoni is in short as follows: after sexual reproduction of adult worms within the tributaries of the inferior mesenteric vein of humans, representing the definite hosts, females produce eggs, characterized by a typical lateral spine. Some of the eggs pass through the vessel wall into the intestine, then reaching the external world with the faeces. Contact with water induces the release of miracidia, which infect freshwater snails. Within these intermediate hosts, thousands of fork-tailed cercariae develop. Losing their tails, they quickly penetrate human skin, pass through the tissue as schistosomula and develop to male or female schistosomes. Clinical schistosomiasis mainly occurs in heavily infected individuals and is caused by egg deposition in the bowel, liver and lung, leading to a chronic granulomatous inflammation and fibrosis.

The envelope consists of Reynolds' layer (R), the inner layer with extreme electron density, the middle layer with intermediate electron density and the outer layer (S), the middle layer with intermediate electron density and the outer layer (S). The envelope consists of Reynolds' layer (R), the von Lichtenberg envelope (Li) and a fluid-filled cavity, Lehmann's lacuna (Le).

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Prof. Hans Christian Korting
Dermatologische Klinik und Poliklinik der Ludwig-Maximilians-Universität München
Frauenlobstrasse 9–11
D-80337 Munich (Germany)
Tel. +49 89 5160 4615, Fax +49 89 5160 4602

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FIG. 1. Paragenital Schistosoma granuloma in the right groin.
FIG. 2. S. mansoni egg surrounded by an infiltrate of eosinophils, lymphocytes and plasma cells.
FIG. 3. S. mansoni egg in the dermis. Magnification × 12,500.
FIG. 4. Composition of egg shell and envelope of mature S. mansoni egg in detail. Parts of the shell are the peripheral microspines of the outer layer (S), the middle layer with intermediate electron density and the inner layer with extreme electron density.

Magnification × 29,000. Bar = 1.0 μm.
tail the typical composition of the egg shell and the envelope of a mature *S. mansoni* egg near the pole. The shell is composed of three layers: the peripheral microspine layer, a middle layer with intermediate electron density and an inner layer with extreme electron density. The space between the developing miracidium and this inner electron-dense layer of the egg shell consists of three parts: Raynolds’ layer, which lies immediately subjacent to the egg shell, the von Lichtenberg envelope, which consists of a single layer of flattened cells, and a fluid-filled cavity, Lehmann’s lacuna, which surrounds the developing miracidium (not shown). The enzyme-linked immunosorbent assay (ELISA) against soluble egg antigen was positive, repeated stool and centrifuged urine examinations for eggs were negative. Abdominal sonography was normal. The patient was diagnosed as having bilharziasis cutanea tarda. Specific treatment was performed by peroral praziquantel (2 × 1.8 g) for 1 day.

**Discussion**

Late cutaneous bilharziasis is a very rare manifestation of the most important human helminthic disease worldwide, which is schistosomiasis.

Originally this mammalian blood fluke infection was an African disease, later exported by the slave trade to Brazil, Venezuela and the Caribbean Islands [1]. Far from endemic areas, travelling habits in modern times constitute a diagnostic challenge for dermatologists and dermatohistopathologists in particular. The deposition of *S. mansoni* eggs in the dermis is mainly associated with severe systemic invasion. Two types of lesions are distinguished, according to their location. The most common sites are the genital and perigenital areas (anus, perineum and groins). *Schistosoma* eggs reach this region by portosystemic anastomoses, presumably via the communication between the inferior mesenteric vein and the rectal venous plexus [2]. The second type, summarized as extragenital lesions mainly occurring on the trunk, is far less common than genital and paragenital lesions and the way of oviposition is not known [3–7]. Polyploid or vegetant tumours, papules or ulcerations are most common manifestations of both types of late cutaneous schistosomiasis. In the very rare cases of isolated skin manifestation without visceral disease, as presented by our patient, lesions are not easily diagnosed by clinical examination. Identification of schistosomal eggs is mandatory for the diagnosis of cutaneous bilharziasis. Although they are easily detected by the histopathologist at an early stage, eggs in late stages are rare and analysis of multiple layers of tissue is necessary to find eggs among the dense connective tissue. Granuloma formation is caused by a delayed-type T-cell-mediated hypersensitivity reaction against soluble egg antigens produced by miracidia and penetrating the eggshell [8]. The ectopic form of this parasitic disease seems to be relatively rare in tropical and subtropical regions with high endemicity probably due to natural immunity depending on genetic factors [7, 9]. Accordingly, ectopic forms of schistosomiasis have been reported more frequently in Europeans visiting the endemic areas for a short time [10–12]. Besides bilharziasis cutanea tarda, cutaneous schistosomiasis and ‘Katayama fever’ are more common cutaneous manifestations of schistosomiasis. An itchy maculopapular rash after repeated exposure to cercariae is well-known hypersensitivity skin reaction also named ‘swimmer’s itch’ [13]. However, this reaction is mainly induced by avian cercariae penetrating human skin without further progression of their life cycle. The dermatitis is more intense than in cases of human schistosomal cercarial penetration and can occur in less than 15 min in sensitized humans.

Urticarial eruption, combined with acute febrile illness, eosinophilia, headache, diarrhoea and cough are typical signs of the ‘acute toxemic’ schistosomiasis or ‘Katayama syndrome’. This second cutaneous manifestation of schistosomiasis is thought to be immune complex mediated and appears suddenly 2–3 weeks after infection.

We here report the very rare case of a patient presenting isolated skin manifestations without any preceding manifestation involving the genitourinary or intestinal systems. Clinicians should be aware of this differential diagnosis even if the patient has returned from endemic areas months before the clinical manifestation. Finally identification of eggs in the dense connective tissue may present problems to dermatopathologists caused by the eggs’ scarcity in the very late lesions.

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**References**