Ectopic Cutaneous *Schistosomiasis mansoni* in the Sacral Region

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Key Words

Cutaneous schistosomiasis · Ectopic schistosomiasis · *Schistosomiasis mansoni* · Late cutaneous schistosomiasis

Abstract

The authors report one case of late cutaneous *Schistosomiasis mansoni* in a biopsy of a skin lesion in the sacral region in a 51-year-old female living in Contagem, Minas Gerais. The patient was treated successfully with oxamniquine (Mansil®).

Introduction

Cutaneous lesions are rare in both acute and chronic forms of schistosomiasis, even in regions where this parasitosis is highly endemic. In Brazil, 31 cases of cutaneous lesions of *Schistosomiasis mansoni* [10] were reported before 1997, mainly in the states of Bahia and Minas Gerais. They were predominantly in extragenital sites, particularly on the anterior chest wall and the abdomen, where vegetating, verrucous, smooth or sometimes ulcerated lesions were formed, with or without sinuous fistulae, either skin-colored or erythematous, asymptomatic or pruriginous, sometimes with a herpetiform or zosteriform distribution [4, 7, 9–11, 20, 21]. However, some cases also occurred in genital [6, 8, 12] and perigenital [22] areas.

Such lesions are often found in biopsy and frequently show eggs and granulomas. Worms are rarely found, but should be viewed seriously when they are [1, 2].

In a review of 4,416 autopsies by Lima [16], no schistosomiasis cases showed ectopic skin lesions. Ectopic lesions are considered to be those located outside the hepatic-intestinal area in accordance with Lima [16].

Case Report

On June 23rd 2008, an incisional biopsy was performed on the sacral lesion of a 51-year-old female patient. Macroscopically, it was an irregular skin fragment, brownish in color, measuring 1.3×0.6 cm. The material was first fixed in formaldehyde, then cut in half, and after washing in alcohol, xylol and liquid paraffin, it was embedded in paraffin and sectioned with a microtome. Sections 3 to 4 micrometers thick were stained with hematoxylin and eosin (HE).

Histopathological examination showed 28 *S. mansoni* eggs (the fragment was thoroughly examined at 3 levels of magnification: 40×, 100× and 400×). Most of them were calcified or represented by empty shells or by remains of miracidia, some with a clear lateral spicule. They were mainly distributed in the reticular dermis, with a few in the papillary dermis and in the basal membrane of the epithelium. There were also several granulomas, well or badly defined, in the production phase. These latter were characterized by giant Langhans cells or the foreign-body type, involving the egg or its shell centrally, surrounded by mononuclear cells with moderate eosinophilia. An intense diffuse inflammatory infiltrate coexisted, predominantly comprising mononuclear cells and a moderate number of eosinophils, involving calcified eggs and egg shells, in the superficial, medium and deep reticular and papillary dermis, unlikely to form granulomas (fig. 1a–d).

Two egg remnants were identified in the Malpighian layer of the epidermis, one a shell and the other a calcified egg, inside tunnels made by the parasites with no involving reaction and with no morphological changes in the epithelial cells (fig. 2a–d).

In addition to these lesions linked to the schistosomiasis infection, there were other independent lesions not directly linked to the parasitosis: irregular acanthosis and papillomatosis, hyper- and moderate parakeratosis and an associated junctional nevus-like lesion were noted.

Diagnosis

The diagnosis was cutaneous schistosomiasis, with granulomas in the production phase and several empty shells and calcified eggs, associated with junctional nevus. In view of the anatomopathological findings, the patient was treated with a capsule containing 250 mg of oxamniquine (Mansil®) daily for 3 consecutive days. There was significant reduction of the lesions, up to complete disappearance with no remains. The patient has since shown good general status, is healthy, flushed, hydrated, anicteric and acyanotic. She presents no edemas, cutaneous lesions and/or complaints regarding the gastrointestinal and cardiopulmonary systems. Among previous examinations, an abdominal ultrasound performed in 2003 is relevant because of a genitourinary tract complaint. The ultrasound detected a nodular image in the right lobe of the liver (casual finding). In view of that image, a computerized tomography was required, dated January 13th 2003. It revealed a lesion compatible with hepatic hemangioma. Liver and spleen showed smooth surfaces with no signs of the hepatosplenic form of schistosomiasis.

Discussion

We have described a case of *S. mansoni* in the sacral region that was chronic cutaneous and also ectopic, found at random during a routine gynecological examination of a woman from the town of Ibiporã (PR), who had lived in the rural area of Coluna (MG) until she was 9 years old and then in the neighboring town of São José do Jacuri (MG), where she remained until she was 20 years old. The patient reported that in both these towns of the Vale do Rio Doce, bathed by the Rio Suaçuí, she was in contact with natural water, and also that several of her relatives used Mansil. When she was 21 years old, she moved to Contagem (MG), where she continued to have frequent contact with water from the Vargem das Flores reservoir, located between the cities of Contagem and Betim. She reported no contact with natural water for about 20 years.

There is an important fact in this previous history: there was a likelihood of reinfection, possibly by different strains, in Coluna and in São José do Jacuri, and also in Contagem, the last place where she had contact with natural water 20 years ago.

Data from the Secretaria de Saúde de Minas Gerais (secretariat of health of the state of Minas Gerais) indicate a high level of infection in Coluna (21.2%) and in São José do Jacuri (55%), but a low index (<10%) in the Vargem das Flores reservoir [24] in Contagem.

We should also mention that on the 5th of April 1992, a group of 24 planorbidae was collected from the Vargem das Flores reservoir. Twenty-one of these showed characteristics of *Biomphalaria occidentalis* as described by Paraense [19]; there were also 3 empty shells, 1 of which was typical of *Biomphalaria glabrata*. A cercarial shedding test on light exposure was negative [17, 19]. According to the authors of the aforementioned research, a single empty *B. glabrata* shell among *B. occidentalis* shells in the place studied indicates ‘competitive exclusion’ between the 2 species, with the advantage going to *B. occidentalis*, which may have been the controlling species in contrast to *B. glabrata*.

B. occidentalis is likely to have been introduced into the reservoir together with the fish supply in 1985, including species from the Rio Paraná, where *B. occidentalis* is found. On the other hand, studies performed before 1992 [15, 24] showed that only *B. glabrata* was present in the Vargem das Flores reservoir when our patient bathed there.

The mean life span of schistosomes in humans is from 3 to 5 years. Nevertheless, it is known that they may survive and produce viable eggs for more than 30 years [3]. Therefore, taking into account that the last contact with natural water occurred about 20 years ago, the worms may have had a long life in the host’s blood vessels in this case. Such cases have been reported in the scientific literature, particularly among tourists [7, 14, 18].

We were unable to determine the clinical form of this parasitosis precisely. The patient was asymptomatic, probably with an intestinal parasite. Ultrasound and computer tomography images allowed us to reject the hypothesis of a Symmers-Bogliolo [23] form.

In addition to the facts already mentioned, other reasons justifying publication of this case are:

- (1) the ectopic location in the sacral region, not described previously;
- (2) the phenomenon of egg extrusion through tunnels in the superficial dermis layer and among epidermis cells with no eosinophilic reaction, which suggests inverse tunnels built from the outside to the inside by cercariae penetrating the human skin [5, 26] – this seems to be novel;
- (3) several *S. mansoni* eggs were observed, many of them calcified, but few induced a granulomatous reaction in their immediate neighborhood. This contrasts with most cases published, in which few eggs and few granulomas were found, several in the initial phase of evolution, i.e. in the necrotic-exudative phase [23]. There was also an intense inflammatory reaction, irrespective of the granulomatous lesion, with relatively few eosinophils, which was diffuse in the superficial, mean and deep papillary and reticular dermis;
- (4) the association of the schistosomiasis with junctional nevus, irrespective of the cause-effect relationship (Raso P).

Finally, we should discuss which of the mechanisms mentioned in the literature [1, 6, 10, 13, 22] may have been involved in the eggs reaching the patient’s skin. In our opinion, the arrival of the worms and the egg deposition in the skin covering the sacral region probably occurred through Batson’s vertebral venous plexus, which shows connections among the pelvic veins and cranial venous sinuses, via the vertebral venous plexus and

superior and inferior venae cavae. It is possible that the worm reached the external vertebral venous plexus surrounding the spinal cord (extrarachidian venous plexus) by this route. The plexus is composed of a network of avascular veins with very thin walls, lacking muscle fibers and easily distended, and it completely covers the spinal cord. Blood from the abdominal cavity may be redirected to this venous plexus in cases of increased abdominal pressure, which may be induced by coughing, defecation, and obesity among other causes. This route might also be responsible for exceptional cases in which the central nervous system and cutaneous lesions are involved simultaneously [20, 27].

Fig. 1. Skin section (sacral lesion). **a** An intense chronic and diffuse inflammatory reaction can be observed, whereas the exudate is predominantly constituted by mononuclear cells involving calcified eggs and egg shells, in the dermis (arrows). Bars = 32 μ m. **b** Higher magnification showing some eosinophils (arrowheads) and a detail of two *S. mansoni* eggs in the center of the figure. Bars = 16 μ m. **c** Note a giant cell formation (arrowhead) engulfing a viable *S. mansoni* egg (arrow). Bars = 32 μ m. **d** A calcified egg can be noted at the left corner of the figure. Bars = 16 μ m.

Fig. 2. Skin section (sacral lesion). **a** Observe two *S. mansoni* eggs where they are identified in the Malpighian layer of the epidermis: one represented by a shell (large arrow) and the other by a calcified egg (small arrow). Bars = 32 μ m. **b** Higher magnification showing a detail of the egg with remains of miracidia into the epidermic layer. Bars = 16 μ m. **c** Note an intense chronic inflammatory reaction in the dermis associated to an epidermic lesions as acanthosis and papillomatosis, hyper- and moderate parakeratosis and the association of junctional nevus-like lesion (arrows). Bars = 32 μ m. **d** Tunnels made by parasites with no inflammatory reaction and with no morphological changes in epithelial cells can be observed. Bars = 16 μ m.

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