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CASE REPORT

Cutaneous ectopic schistosomiasis associated with Löffler syndrome. A rare case report

Esquistossomose ectópica cutânea associada a síndrome de Löffler. Um raro relato de caso

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Abstract

Schistosomiasis mansoni is an endemic disease in Brazil, usually causing systemic symptoms, mainly gastrointestinal. Skin lesions are best described in the acute phase of the infection, with ectopic skin lesions rarely seen. We report a case of schistosomiasis with rare ectopic cutaneous involvement, with perianal papules and plaques, acquired in the state of São Paulo in 2020. At the beginning of the investigation, the hypothesis of infection by COVID-19 was raised, due to a pulmonary condition, but it was discarded after a negative PCR for the virus. Due to eosinophilia, pulmonary CT characteristics and epidemiology, Löffler's syndrome was suspected. The definitive diagnosis of schistosomiasis was given, after the onset of the skin rash, by the anatomopathological examination of the skin biopsy, thus avoiding an invasive examination—lung biopsy, in the patient who was using anticoagulants. The protoparasitological examination of faeces was negative. The patient was treated with Praziquantel, with the improvement of the condition. This report demonstrates the importance of dermatological examination and skin biopsy for the definitive diagnosis of Schistosomiasis in a patient with severe systemic manifestations.

Keywords: Cutaneous schistosomiasis. Schistosomiasis mansoni. Neglected diseases. Parasitic diseases. Loffler syndrome.

Resumo

A esquistossomose mansônica é uma doença endêmica no Brasil, causando geralmente sintomas sistêmicos, principalmente gastrointestinais. As lesões cutâneas na doença são melhores descritas na fase aguda da infecção, sendo que lesões cutâneas ectópicas são raramente vistas. Relata-se o caso de esquistossomose com acometimento cutâneo ectópico raro, localizado à região perianal e glútea, adquirido no estado de São Paulo em 2020. No início da investigação diagnóstica fora aventada hipótese de infecção por COVID-19, devido quadro pulmonar, mas esta foi descartada após PCR negativo para o vírus. Em decorrência da eosinofilia, características da TC pulmonar e epidemiologia suspeitou-se de síndrome de Löffler. O diagnóstico definitivo de esquistossomose foi dado, após início do quadro cutâneo, pelo exame anatomopatológico da biopsia

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da lesão, evitando assim um exame invasivo - biópsia pulmonar, no paciente que estava em uso de anticoagulantes. O exame protoparasitológico de fezes foi negativo. O paciente foi tratado com praziquantel, evoluindo com melhora do quadro. Este relato demonstra a importância do exame dermatológico e da biópsia de pele para o diagnóstico definitivo num paciente com manifestações sistêmicas graves.

Palavras-chave: Esquistossomose cutânea. Esquistossomose mansoni. Doenças negligenciadas. parasitose. Síndrome de Loffler.

Introduction

Schistosomiasis mansoni is a systemic disease caused by a trematode helminthic which represents a public health problem in Brazil and in the world[1].

Skin lesions are observed mostly in the acute phase (swimmer`s itch or cercarial dermatitis), and are very unusual in the chronic forms of schistosomiasis, even in endemic areas[2].

The cutaneous form is usually asymptomatic, and it occurs often in white young women. Chronic lesions are formed when the eggs or worms migrate to the skin, causing granulomas on the skin and mucous membranes[3,4].

We report the case of a male patient, who after a trip to the coast of São Paulo, developed the systemic schistosomiasis that was correctly diagnosed only after skin biopsy.

Case report

A previously healthy male patient–D.G.D., aged 17-year-old, used to swim in river waters on family trips. One month after his trip to "Juréia" and "Boracéia," in the South Coast of São Paulo state, in February 2020, he developed fever, dry cough, and diarrhea, progressing with prostration and desaturation, requiring hospital admission and additional oxigenotherapy. With the hypothesis of viral infection (COVID-19 or Influenza), he was medicated with Oseltamivir and Amoxicillin Clavulanate, but PCR for COVID-19 and Influenza were negative.

A computed tomography (CT) of the thorax revealed multiple solid sparse nodules distributed bilaterally on the lung parenchyma, some of them with halo in opaque glass, measuring up to 10 mm, and additional splenomegaly. Laboratory examinations showed leucocytosis (15.090/mm^3) with 38.2% eosinophils (5.760/mm^3). A protoparasitological examination of feces was negative. Pneumonology suspected of Löffler`s Syndrome, and prescribed Ivermectin 18 mg/day for 2 days and Albendozole 400 mg/day for 6 days, aiming to treat *Ascaris lumbricoides* or *Strongyloides stercoralis*. Nevertheless, one day after onset of this treatment, he developed a cutaneous rash on the dorsum, axillae and inguinal region (Fig. 1), suggesting possible toxemia secondary to antigen release from the dead helminths. Hydrocortisone 100 mg every 12 hours was soon initiated with improvement of the rash in a few days.

However, just 2 days after stopping Ivermectin, the patient developed symmetrical erythematous papular lesions, some isolated and with minor excoriations and others confluent into small plaques, localized on the buttocks and perianal area (Fig. 2). A skin biopsy of the right buttock demonstrated eosinophilic granulomas associated with typical parasitic eggs (Fig. 3), confirming the diagnosis of Schistosomiasis. Also indirect immunofluorescence assay to *S. mansoni* was positive 1/64. Treatment with praziquantel, 4.2 g as a single dose induced total relief, improving his clinical status after 1 week, including complete resolution of perianal cutaneous lesions.

Discussion

Schistosomiasis is still a serious health problem in Brazil[6], with *S. mansoni* as the only species found in this country[8]. It is acquired in contact with contaminated water, and transmission depends on snails of the genus Biomphalaria, found in freshwater-bathed regions[7]. When carrying out their domestic and leisure activities in rivers, lakes and ponds, humans are exposed to the larva, that have left the snails, which actively penetrate the skin and mucous membranes[7]. The larvae reach the blood vessels and portal circulation, where they will become adult worms[8].

Soon, after the infective contact, cercarial dermatitis may occur, which presents with pruritus, usually transient, and an erythematous micropapular eruption. After about 40-60 days, coincident with the laying of eggs[6], usually in the mucosa and submucosa of the colon and rectum veins[9], patients develop high fever, chills, sweating, malaise and asthenia, followed by diarrhea, nausea, and vomiting, which constitute the phase entitled Katayama fever. Associated

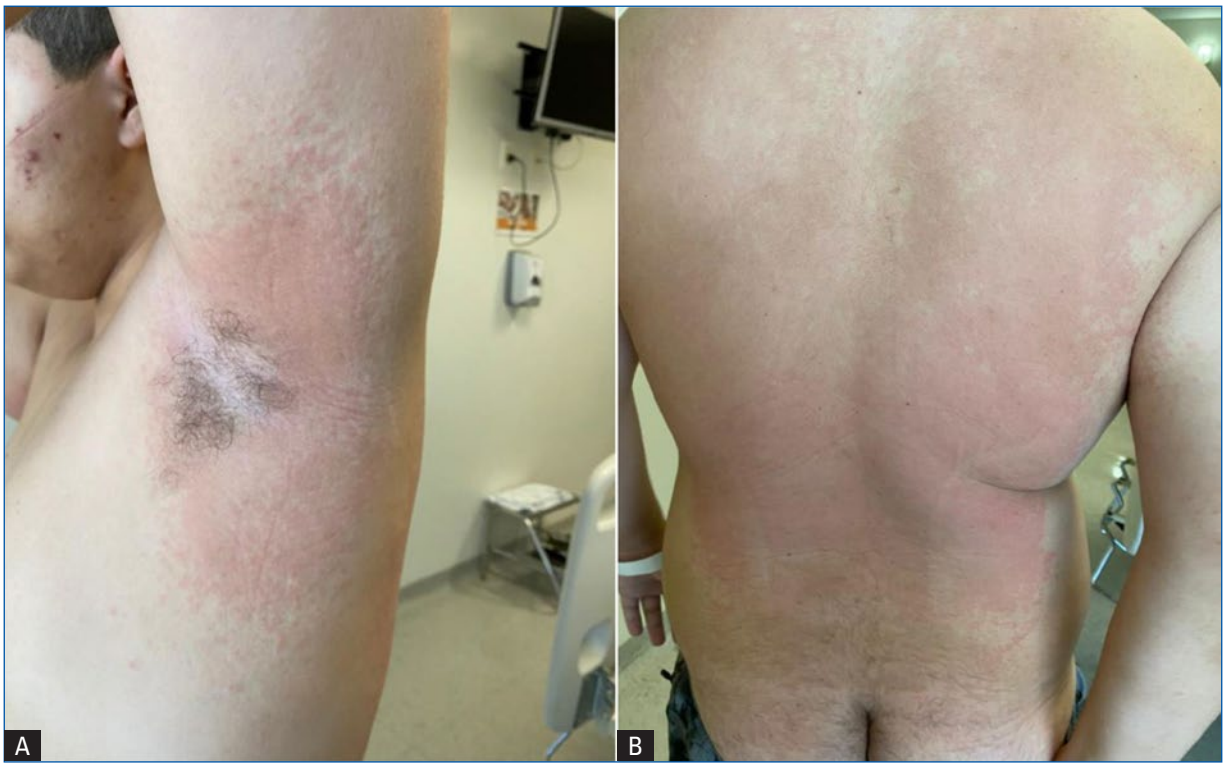

Figure 1. A and B: maculopapular exanthema with predominant involvement of the axillae (**A**), inguinal region and dorsum (**B**), which developed within 24hours on onset of Albendazole and Ivermectin.

hepatosplenomegaly and significant eosinophilia, often lead to the diagnosis of acute schistosomiasis[7]. In its chronic phase that occurs more than 6 months after infection various organs may be affected with signs and symptoms of intestinal, hepato-intestinal or hepatosplenic involvement, as well as neurological and cutaneous forms[7,8]. Tissue granulomas may form in response to the presence of the eggs, classically leading to bowel wall thickening, periportal fibrosis of the liver and portal or pulmonary hypertension[10].

Pulmonary involvement can be divided into two vascular clinical forms: hypertensive and cyanotic. The first is characterized by dyspnea on exertion, palpitations, dry cough, and constrictive chest pain, which can also lead to extreme asthenia and fatigue, in addition to heart failure. The cyanotic form has a worse prognosis and presents with generally mild cyanosis, especially in the extremities[6]. Pulmonary manifestations can also occur during the migration of larval forms, such as the so-called Loffler syndrome, manifesting with dyspnea and dry cough and characterized by accumulation of eosinophils in the lungs[10], which is consistent with the initial presentation of the present case that did not show the typical signs of Katayama fever.

Skin lesions of chronic schistosomiasis are generally characterized as papular, ulcerative, granulomatous, and fistulous lesions, often affecting the skin of the genital and perianal region, secondary to the deposition of eggs contiguous to the pelvic vessels[9]. Infiltrative lesions in these regions can cause an inflammatory and fibrotic reaction, leading to the formation of granulomas that begin as asymptomatic firm papules that grow slowly, until they become vegetative[8]. In rare circumstances, the eggs are lodged in extragenital skin, causing cutaneous ectopic schistosomiasis[9], and lesions may be found on the back and abdomen. The exact mechanism of egg deposition in the skin remains unknown, suggesting that anastomoses between venous systems would be associated with the migration of eggs or adult worms to ectopic sites[8].

Diagnosis is based on characteristic lesions, compatible epidemiology and histopathological examination[3]. Histopathology provides identification of *S. mansoni* eggs, surrounded by inflammatory cells, mainly lymphocytes and eosinophils in more recent lesions, but in old lesions necrosis and granulomatous infiltrates predominate[2,10]. The coprologic examination can be used preferably with the use of quantitative

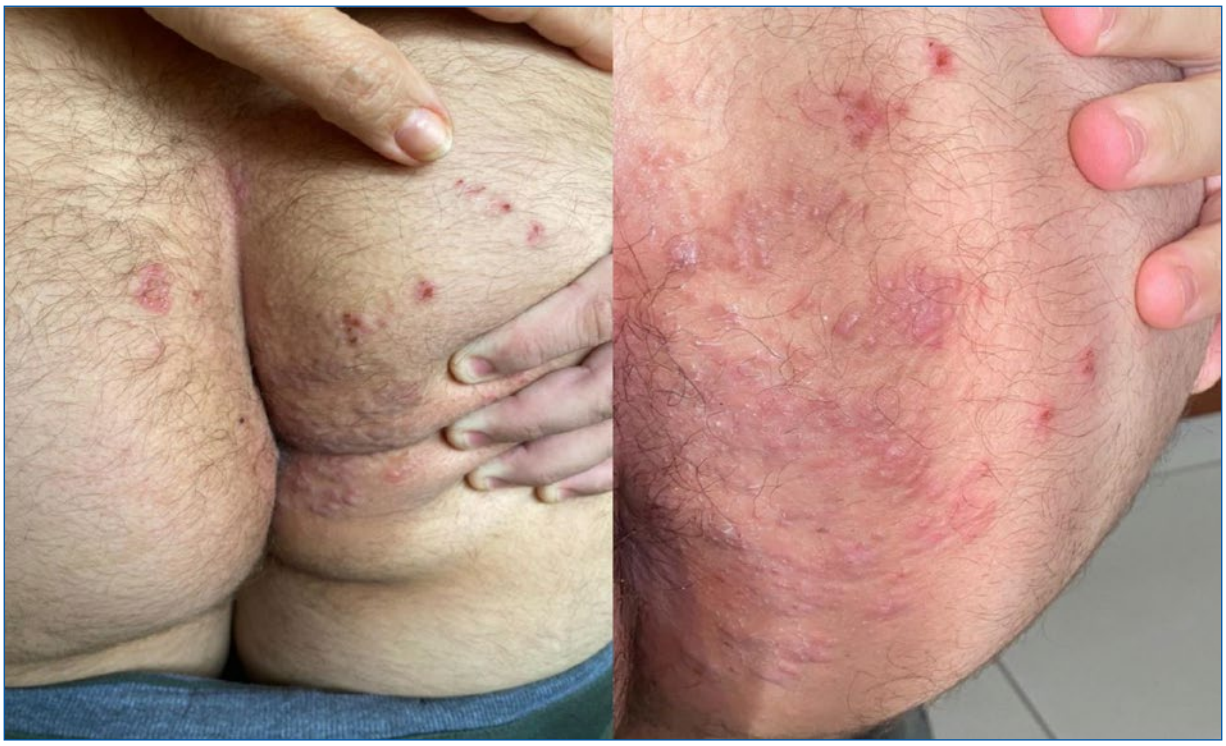

Figure 2. Erythematous papules some isolated, others grouped in small plaques, others ulcerated on the buttocks.

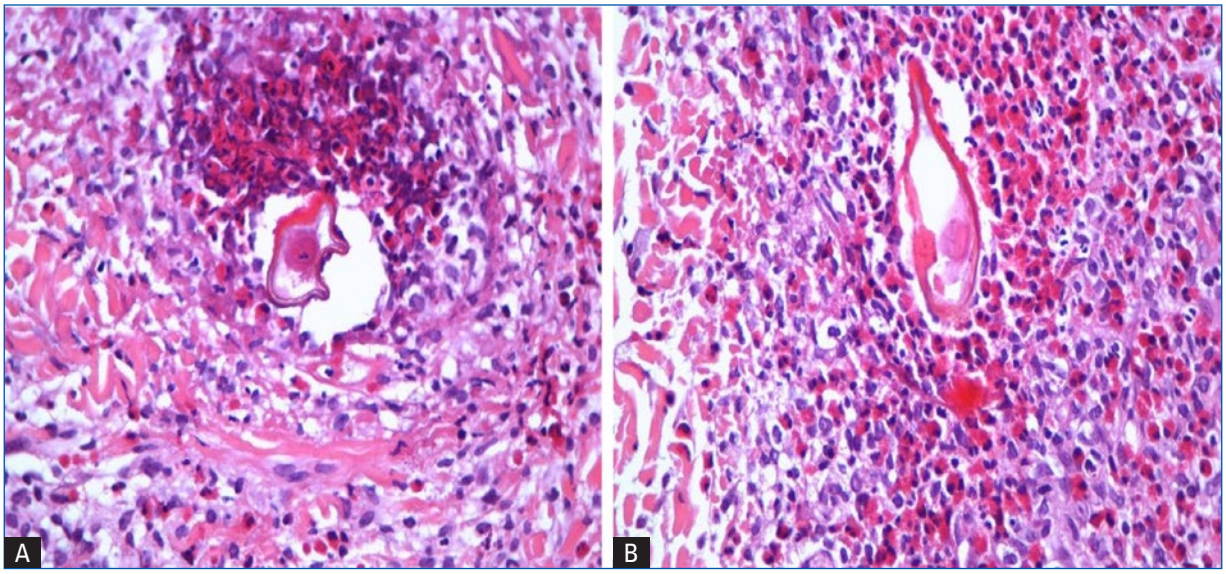

Figure 3. A and B: Histopathological images of the skin biopsy, showing of typical egg of Schistosoma mansoni, surrounded by a granulomatous reaction very rich in eosinophils (H&E–300x).

and sedimentation techniques[4]. Diagnosis can also be confirmed by combining the results of an indirect hemagglutination assay and enzyme-linked immunosorbent assay test, which reach a sensitivity and specificity beyond 90 and 97% respectively[5]. As in our case significant eosinophilia is very frequent. Meltzer et al. described that 53.7% if patients with significant eosinophilia after a trip were diagnosed with schistosomiasis. Around 47.7% of the patients suffered from schistosomiasis acute crisis, 16.9% patients presented chronic symptoms and 36.6% were asymptomatic[5].

The gold standard treatment is praziquantel 60 mg/kg for children until 15 years old and 50 mg/kg for adults, in a single dose with high healing rates (60-90% in endemic areas and about 100% in non-endemic areas)[2,10]. The second-choice drug is the oxamniquine 15 mg/kg, a single dose in adults, and 20 mg/kg, as single dose on children up to 15 years old.

What does this study add to the current knowledge?

This study approaches a rare case of cutaneous schistosomiasis with Löffer syndrome, a rare presentation, despite being an endemic disease in Brazil. We`ve alerted to the necessity for the accurate diagnosis, which in this case was based on the skin biopsy.

Acknowledgments

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Ethical considerations

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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