CASE REPORT

An Atypical Presentation of Acute Schistosomiasis

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Abstract

Schistosomiasis is usually an imported, rare disease in developed countries. The schistosome life cycle has three phases in the human host, each one usually associated with different clinical symptoms. We report a case of a male adolescent presenting with periorbital and facial edema, with no other symptoms or findings during physical examination. Laboratory investigation revealed hypereosinophilia. A thorough medical history revealed that he traveled through several African countries and participated in freshwater activities the month before. A serologic test performed for *Schistosoma mansoni* was positive. He was successfully treated with praziquantel.

We intend to highlight the challenges of a schistosomiasis diagnosis and to present, to our knowledge, the first pediatric case presenting with angioedema. An exhaustive anamnesis and the finding of eosinophilia were fundamental for raising suspicion. Serologic tests may be necessary for diagnosis since a microscopic test has low sensitivity. Treatment with praziquantel may have prevented the progression of the disease and further complications.

Keywords: Adolescent; Angioedema/etiology; Eosinophilia/etiology; Portugal; Praziquantel/therapeutic use; Schistosoma mansoni; Schistosomiasis mansoni/diagnosis; Schistosomiasis mansoni/drug therapy

Introduction

Schistosomiasis is a parasitic disease caused by some species of blood trematodes belonging to the genus *Schistosoma*.[1,2] Three major species infect humans: *Schistosoma haematobium, Schistosoma japonicum*, and *Schistosoma mansoni*.[1-3]

Humans may acquire the infection through skin lesions or mucosa, when coming into contact with cercaria contaminated water.[1-6] The schistosomula migrate via venous circulation to the lungs, then to the heart, and finally develop in the liver, exiting it via the portal vein system when mature.[1,2,6,7] One to three months later, the female worm deposits its eggs in the small venules of the mesenteric or perivesical systems, and the eggs are moved progressively toward the lumen of the intestine (*Schistosoma mansoni*, *Schistosoma japonicum*) or of the bladder and ureters (*Schistosoma haematobium*), being excreted through feces or urine, respectively.[1,2,7] Parasite eggs may become trapped in the host tissues during migration or may be embolized to the liver, spleen, lungs, or cerebrospinal system.[1-3,6,7]

Clinical manifestations vary widely within each infectious phase and can be highly unspecific. In the first phase, percutaneous penetration of cercaria causes a temporary localized dermatitis, typically on the feet or lower legs, called the 'swimmer itch', lasting for a few hours to days.[2,5-8] The second phase (within three to eight weeks), called acute schistosomiasis syndrome or Katayama fever, is associated with a transient hypersensitivity state with unspecific mild symptoms such as fever, myalgia, arthralgia, dry cough, diarrhea, abdominal pain, urticaria, angioedema, and headache.[1-3,6,9] In the third or chronic phase (approximately eight weeks after infection), clinical manifestations vary according to organ involvement, intensity of infection and individual immune response.[2,3] Ectopic schistosomiases can lead to unexpected morbidities such as neuroschistosomiasis, one of the most severe clinical outcome.[3,10]

We report a case of a male adolescent with facial edema and eosinophilia probably due to a *Schistosoma mansoni* infection.

Case Report

A previously healthy black 14-year-old boy born in Angola, living in Portugal for the past four years, presented to the pediatric emergency department,

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a week after returning to Portugal from holidays in several African countries. He presented with large, pale, non-pitting, periorbital and facial subcutaneous edema, headache, and myalgia since the previous day, with no other concomitant symptoms such as fever. The clinical examination was unremarkable, except for the periorbital and facial edema and he had normal blood pressure. Blood analysis showed a normal red and white blood cell count as well as a negative C-reactive protein and a negative heterophile antibody test. Urine analysis revealed the absence of blood or proteins. Presuming a viral infection, he was discharged with symptomatic measures, only to return seven days later with the same symptoms. Once again, clinical evaluation revealed no other abnormal signs, blood analysis showed once again normal ranged red cells and total leucocyte count, but a mild peripheral hypereosinophilia of 2,750 cells/µL. Renal function, electrolytes, liver function, C-reactive protein and urinalysis were all normal. He was then referred to a pediatric outpatient clinical appointment for further investigation.

At this medical appointment, one week later, he was asymptomatic, presenting no abnormal findings on physical or neurologic examination, although multiple punctate cicatricial lesions on the lower limbs were observed, with no signs of recent interdigital lesions. After exhausting anamnesis, he admitted traveling to several African countries on vacation the previous month, where he swam in lakes and practiced spearfishing.

Blood tests were repeated on that same day revealing an eosinophilic count of 4,100 cells/µL, with a normal leucocyte count of 9,620 cells/µL. The other blood and urine analysis remained normal. Serologic testing for *Schistosoma mansoni* was positive (immunoglobulin G), although no parasite eggs were detected in the stool. Serologic tests for cytomegalovirus, toxoplasmosis and Epstein-Barr virus showed no acute infection, and coinfection with other parasitic diseases, such as *Toxocara canis*, filaria and *Strongyloides stercoralis* were excluded. Normal urinalysis, abdominal ultrasound, cardiac ultrasound, and electrocardiogram excluded the involvement of other organs.

He was treated with a single dose of praziquantel 40 mg/kg, with the total remission of the clinical and laboratory findings. There was a gradual decrease of the blood eosinophil count to a normal value (360 cells/µL) six months after praziquantel, and a microscopic examination of urine and stool still revealed no evidence of parasite eggs. He remained asymptomatic after eight months of follow-up.

Discussion

This clinical report illustrates the main challenges of a schistosomiasis diagnosis, in a developed non-endemic country. Although schistosomiasis is widespread throughout developing countries, it is a rare disease in Portugal and only usually seen in travelers or immigrants. Schistosomiasis may be asymptomatic or present with unspecific symptoms, as observed in this case. Presentation with angioedema, although rare, may be a part of cutaneous manifestations of the acute schistosomiasis syndrome. Skin involvement occurs with highly variable frequency and intensity, and it is described in 12% of adults with schistosomiasis due to *Schistosoma mansoni*.[8,11] To our knowledge, this is the first pediatric report of such a presentation.

It should be emphasized that symptoms can persist up to three months after infection.[2,5,8] An exhaustive anamnesis, including travel history and aquatic recreational activities in endemic areas, should be conducted even when presenting symptoms do not immediately suggest a tropical etiology.

Differential diagnosis of facial edema should include local infections such as orbital cellulitis, odontogenic infection, and sinusitis as well as systemic diseases such as hypothyroidism, Epstein-Barr virus infection, and nephrotic syndrome.[12]

Eosinophilia is a hallmark of parasitic infections, and its degree depends on the stage, intensity, and duration of infection.[2] In the presented case, diagnosis was suggested by the anamnesis associated to the increasing hypereosinophilia, suggesting a recent infection.

The gold standard for the diagnosis of schistosomiasis is the observation of parasite eggs by microscopic examination of urine, stool, or tissue biopsy samples.[5,13] In this case, stool and urine examinations for ova, cyst, and parasites were negative, but as described in the literature, eggs tend to be excreted intermittently. In some cases, the quantity of eggs excreted to reach the detection threshold occurs only 12 weeks after infection and if present in small quantities, may not be detected.[1,13] We considered that our patient parasite load was probably low, and relied the diagnosis on serologic tests. Nonetheless, these tests cannot discriminate exposure from active infection in people from endemic regions.[3,7]

Praziquantel is the first line treatment for all forms of schistosomiasis.[1,3,5,6] It should be preferably started four to six weeks following exposure, when worms are expected to be fully matured.[14] Regarding immature forms, antimalarial drugs have shown to be effective.[15] In our case, as soon as the diagnosis was made, approximately six weeks after presumed exposure,

treatment was instituted. With a single dose of 40 mg/kg, 70%-100% of patients cease to excrete eggs.[2,7] When egg excretion remains, despite adequate treatment, egg counts and antigen concentrations show a reduction of more than 95%, minimizing the risk of disease progression.[5,7]

Due to the possibility of unspecific presenting symptoms, schistosomiasis infection should be considered in all travelers or immigrants from endemic regions. Aquatic activities practice should be inquired directly in order to raise diagnosis suspicion. To reduce the rate of infection, all travelers to endemic countries, including migrants, should be warned of the risks of schistosomiasis and which preventive measures to adopt.

WHAT THIS CASE REPORT ADDS

- In developed countries, schistosomiasis is only usually seen in travelers or immigrants.
- The schistosome life cycle explains different clinical symptoms.
- Angioedema is a rare symptom that has not been previously described in the pediatric population.
- Hypereosinophilia should raise suspicion of a parasitic disease.
- Treatment with praziquantel is crucial to avoid the progression of the disease and further complications such as neuroschistosomiasis.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Uma Apresentação Atípica de Schistosomíase Aguda

Resumo:
A schistosomíase é uma doença rara, habitualmente importada em países desenvolvidos. O ciclo de vida do schistosoma tem três fases no hospedeiro humano, associadas a diferentes sinais e sintomas. Reportamos o caso de um adolescente com edema periorbitário e facial, sem outros sinais ou sintomas. A avaliação laboratorial mostrou eosinofilia. Uma história clínica exaustiva revelou que o adolescente tinha viajado para vários países africanos com prática de atividades aquáticas, no mês prévio. A avaliação serológica para *Schistosoma mansoni* foi positiva. Foi tratado com praziquantel, com remissão dos sintomas.
Pretendemos alertar para o desafio diagnóstico da schistosomíase sendo este, até à data, no nosso conhecimento, o primeiro caso pediátrico que se apresenta com angioedema. Uma anamnese exaustiva e a eosinofilia foram fundamentais na suspeição diagnóstica. A serologia pode ser necessária, pois os testes microscópicos têm baixa sensibilidade. O tratamento com praziquantel poderá ter evitado a progressão e complicações desta doença.

Palavras-Chave: Adolescente; Angioedema/etiologia; Eosinofilia/etiologia Esquistossomose mansoni/diagnóstico; Esquistossomose mansoni/tratamento farmacológico; Portugal; Praziquantel/uso terapêutico; Schistosoma mansoni