Rev. Inst. Med. trop. S. Paulo
40 (4): 253-257, July/August, 1998.

ECTOPIC CUTANEOUS SCHISTOSOMIASIS: REPORT OF TWO CASES AND A REVIEW OF THE LITERATURE

José de Souza ANDRADE FILHO(1), Maria Sueli S. N. LOPES (2), Antônio A. CORGOZINHO FILHO (3) & Gil P. M. PENA (4)

SUMMARY

Two cases of ectopic cutaneous schistosomiasis are described. Both patients presented with abdominal papular skin lesions, which on biopsy were found to contain granulomas with *Schistosoma mansoni* eggs. Twenty-five other cases were retrieved from the literature. Most patients were female, mean age 24.9 year, with a predominance of the white race. The most common localization was anterior thorax and abdomen. Usually, the lesions were asymptomatic. In few cases, however, severe clinical syndromes due to the parasite coexisted, such as transverse myelitis or the acute-toxemic form of the disease. Intestinal infection was not frequently demonstrated in these patients. The importance of the recognition of these cutaneous lesions may rest on the opportunity to provide an etiological diagnosis in these difficult cases.

KEYWORDS: Schistosomiasis; Skin diseases.

INTRODUCTION

Adult *Schistosoma mansoni* worms are usually lodged in venules of the mucosa and submucosa of the colon and rectum. By virtue of this localization, ova are frequently found in the colon and liver. After the development of portal hypertension and the establishment of a portacaval shunt, they can also be found in the lungs.

Despite high endemicity of this parasitosis in several parts of the world, the finding of schistosome eggs or worms in other organs constitutes a rare phenomenon, sometimes with serious clinical complications, usually reported in the literature as pathological curiosities. The possible implications that these ectopic localizations may have on the comprehension of the parasite behavior within host environment has been given little attention.

We have had the opportunity to observe two cases of cutaneous ectopic localization of *Schistosoma mansoni* infection. We stress out the clinical aspects of this rare dermatologic affection, as well as possible implications these observations may have on the understanding of the parasite behavior within host.

CASE REPORT

Case 1

A 14-year-old white boy with history of intermittent fever, diarrhoea (10 evacuations/day), weight loss (about 12 kg in two months), was seen at the hematology section, in March, 1994, because of intense leukocytosis with eosinophilia. He also presented papular cutaneous lesions in periumbilical region bilaterally (Fig. 1), which appeared about 20 days earlier.

In January, 1994, during a bath in Paraopeba river, in the city of Esmeraldas, State of Minas Gerais, he noted cutaneous pruritic lesions in the left foot. In retrospect, these lesions were presumed to represent cercarial dermatitis.

Laboratory studies revealed high erythrocyte sedimentation rate (89 mm/60 min), leukocytosis (WBC = 34100/mm^3), with eosinophilia (25516 eosinophils/mm^3), slight elevation of hepatic enzymes (SGOT = 50 UI/L and SGPT = 58 UI/L - normal range 8-25 UI/L; alkaline phosphatase = 182 UI/L - normal range 12-43 UI/L; GGT = 76 UI/L - normal range 19-24 UI/L).

Stool samples were repeatedly negative for *Schistosoma* eggs. An intradermal test was performed, also with negative result.

Trabalho desenvolvido no Serviço de Anatomia Patológica do Hospital Felício Rocho.
(1) Professor Titular de Patologia, Faculdade de Ciências Médicas de Minas Gerais; Chefe do Serviço de Anatomia Patológica, Hospital Felício Rocho. Belo Horizonte. MG, Brasil.
(2) Serviço de Hematologia do Hospital Felício Rocho. Belo Horizonte, MG, Brasil.
(3) Médico Dermatologista (Clínica Particular). Belo Horizonte, MG, Brasil.
(4) Serviço de Anatomia Patológica do Hospital Felício Rocho. Belo Horizonte, MG, Brasil.
Correspondence to: Dr. Gil P. M. Pena. Rua Uberaba, 418/102. 30180-080 Belo Horizonte, MG, Brasil. Fax 55 31 295-2688. E-mail: gilpena@unix.horizontes.com.br

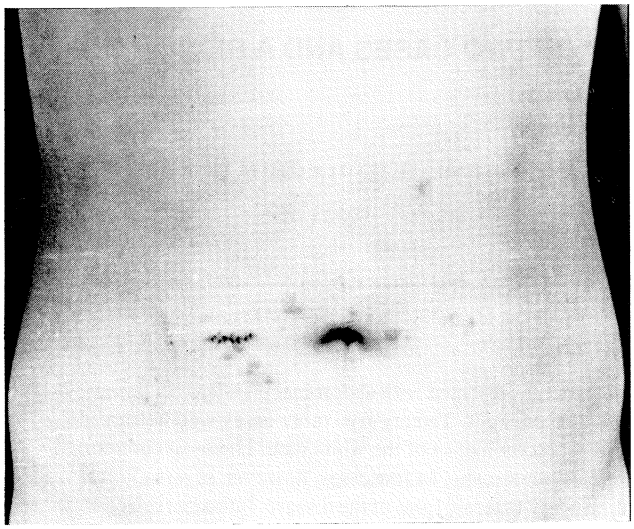

Fig. 1 - Clinical aspect of ectopic cutaneous schistosomiais (case 1). Randomly distributed papular skin lesions in periumbilical area, bilaterally.

An abdominal echography showed hepatosplenomegaly and periaortic and portacaval lymphadenectasis.

Histopathological examination of periumbilical skin lesions revealed granulomas with *Schistosoma mansoni* eggs. (Fig. 2)

Fig. 2 - *Schistosoma mansoni* egg, with characteristic lateral spine in dermis from histopathological examination of periumbilical skin lesions (case 1).

Case 2

A 32-year-old white man presented with dermatologic lesions characterized as small, non-pruritic papules in left abdomen, extending from around umbilicus to left hypocondrium, which appeared three months before (Fig. 3). Any other symptom was denied. No history of previous contact with possibly infected waters could be assured. The patient reports a travel to a river shore, three years before, but denies any direct contact with the water. The lesions were suspected clinically to represent micronodular sarcoidosis, lichen planus or ectopic cutaneous schistosomiasis.

Histopathological picture was characterized by dermal granulomas containing schistosomal eggs, some of them calcified. Stool examination or rectal biopsy was refused by the patient.

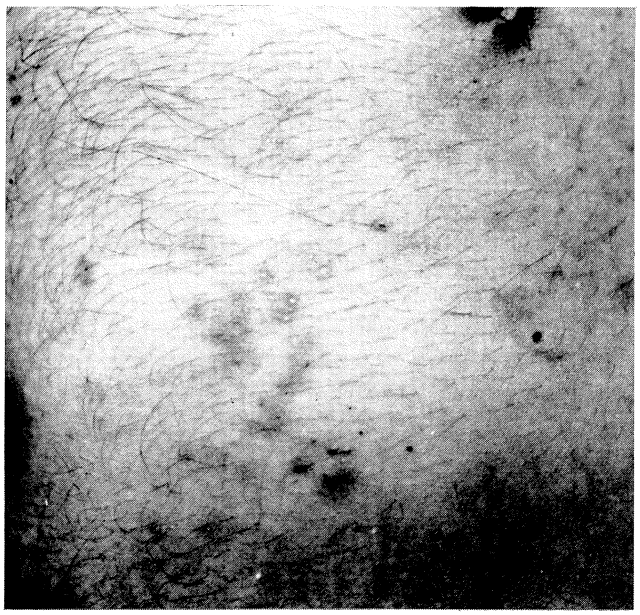

Fig. 3 - Clinical aspect of ectopic cutaneous schistosomiais (case 2). Papular skin lesions in left periumbilical area. Lesions are sometimes confluent with a slightly erythematous halo.

COMMENT

In a review of the literature, 25 cases of ectopic cutaneous schistosomiasis mansoni could be retrieved (Table 1). Main demographic variables and other relevant characteristics are presented in Table 2. The majority of the patients were female with a predominance of the white race.

The majority of lesions presented in the anterior trunk (14/25). This occurrence is anatomically explained by the connections of portal and systemic venous circulation territories. On an analysis of the distribution of trunk lesions, one can easily recognize the patterns of distribution of engorged veins in collateral circulation

TABLE 1
Reported cases of ectopic cutaneous schistosomiasis mansoni.

Case no.	sex	race	age	Estimated time since infection	Intestinal Infection	Procedence	IDR(1)	Localization	Ref.
1	M	white	41	3 mo	yes	S. Africa	NS	Superior thorax (2)	4
2	F	white	24	10 y	no	Venezuela	+	Abdomen, thorax (5)	2
3	F	white	4	3 mo	yes	Brazil (MG)	NS	Foot sole (3,6)	11
4	F	white	26	4 mo	no	Brazil (MG)	NS	Ant. thorax	11
5	F	white	15	3-4 mo	yes	Brazil (MG)	NS	Ant. thorax	11
6	M	NS	33	6 y	no	Africa	NS	Paraspinal area (3)	15
7	F	NS	33	NS	yes	Saudi Arabia	NS	Ant. abdomen	9
8	M	NS	24	5 mo	no	Puerto Rico	+	Periumbilical region	13
9	F	non-white	32	NS	NS	Brazil	NS	post. thorax	12
10	F	white	17	9 y	NS	Brazil	NS	ant. post. thorax	12
11	F	white	26	2 mo	no	Brazil (BA)	NS	lumbar region	1
12	F	white	28	2 mo	no	Brazil (MG)	NS	lumbar region (6)	1
13	NS	NS	15	NS	NS	Brazil	NS	nose (7)	1
14	M	non-white	13	NS	yes	Nigeria	+	head	8
15	NS	NS	NS	NS	NS	Brazil (MG)	NS	thorax	5
16	F	non-white	22	NS	no	Brazil (BA)	+++	cervical/supraclavic	7
17	F	white	33	NS	no	Brazil (BA)	+++	abdomen	7
18	M	white	23	24 mo	no	Brazil (BA)	+++	abdomen (5)	7
19	F	white	30	20 y	no	Brazil (BA)	+++	abdomen (5)	7
20	F	non-white	29	3 mo	no	Brazil (BA)	+	right hipocondrium	6
21	F	non-white	32	NS	no	Brazil (BA)	++	right hipocondrium	6
22	F	white	23	4 mo	no	Brazil (BA)	+++	right hipocondrium	6
23	F	white	29	6 mo	yes	Brazil (MG)	NS	periumbilical	14
24	M	NS	17	1 mo	yes	Tanzania	NS	front (4)	10
25	M	NS	28	3 mo	yes	Brazil (BA)	NS	face	3

(1) Intradermal reaction with Schistosomal antigens.
(2) Presumptive egg embolism to the brain.
(3) transverse mielytis due to schistosomal eggs
(4) schistosomal ocular involvement
(5) migratory lesions
(6) history of previous treatment
(7) Dr. Z. A. Andrade's case, cited by Bittencourt et al. (1979)
M=male; F=female; NS=not stated; mo=months; y=years

TABLE 2
Summary of major demographic and clinical characteristics of reported cases of ectopic cutaneous schistosomiasis mansoni.

Characteristics	Frequency
Sex (M/F)	8/15
Race (white/non-white)	13/5
Age (mean)	24.9
Intestinal infection	7/21
Eosinophilia	6/15
Localization	
anterior trunk	14/25
posterior trunk	4/25
anterior and posterior trunk	1/25
head/neck	4/25
cervical/supraclavicular	1/25
members	1/25
History of previous treatment	2 cases
Migratory pattern of cutaneous lesions	3 cases
Other associated ectopic localization	3 cases

in cases of portal hypertension. This may indicate that worms and eggs might have reached the cutaneous veins through naturally existing connections between systemic and portal venous territories. This distribution favours a previous passage of worms through portal territory before adult worms reach their ectopic oviposition place. In four cases, the lesions were located in the head[1,3,8,10]. In one case, with associated transverse myelitis, the cutaneous lesion was located in the foot sole[11]. In another case, also with associated transverse myelitis, the lesions were distributed in paravertebral area[15]. In three other cases, lesions were also located in posterior trunk[1,12]. In one case, lesions coexisted in both anterior and posterior thorax[12].

The cutaneous lesions were described as papules. Papules were variously characterized as erythematous, clear, firm, slightly keratotic, skin-colored, hyperchromic or shiny. As a rule, the papules tended to coalesce, forming plaques. In one instance, the lesion was characterized as a hypopigmented plaque, with no discernible papule elements[8].

Intestinal infection could only be detected in about one third of the cases, by means of stool examinations or rectal biopsy. However, rectal biopsy was performed in only four patients, and was positive in one, with previous negative stool examinations. Since rectal biopsy is considered more sensitive than stool examinations, a somewhat higher proportion of intestinal infection might be expected.

Incomplete data were available in respect to eosinophil counts of reported cases. In 15 cases, hematological status was stated; in six cases, eosinophilia was reported by the authors[1,4,6,7,14]. In none of them, there were indications of an acute-toxemic form of the disease. The case 1, presented here, seems to be the first one to present the association of these two rare forms of the disease. The dramatic clinical course, together with negative intradermal and stool examinations, has led to the diagnostic hypothesis of a rare form of eosinophilic leukaemia. The skin biopsy, however, provided the opportunity for a prompt diagnosis.

A very interesting aspect of ectopic schistosomiasis is the apparent tendency of these patients to present ectopic localizations in other organs. This association was observed in three cases, in which cutaneous lesions presented in more unusual localizations. In two cases, in which ectopic cutaneous schistosomiasis was associated with medullary involvement, the cutaneous lesions were present in the foot sole[11] and paravertebral area[15]. In another case, the cutaneous lesion, located in the face, was associated with inflammatory ocular schistosomal disease[10]. A presumptive case with egg embolism to the brain was also reported[4].

Other important aspect is the observation of a migratory pattern of cutaneous lesions observed in some cases[2,7]. This may represent an evidence of the migratory activity of adult worms within host. Probably, this migratory activity also occur in its usual habitat, the colon venules. An abnormal migration related to incomplete or unsuccessful treatment does not seem to play a major role in the pathogenesis of ectopic cutaneous schistosomiasis. A history of previous treatment could be retrieved in only two cases[1,11].

RESUMO

Esquistossomose cutânea ectópica: relato de dois casos e revisão da literatura

Dois casos de esquistossomose cutânea ectópica são relatados. Ambos os pacientes apresentavam lesões papulares no abdômen que, ao exame histopatológico, mostraram presença de granulomas contendo ovos de *Schistosoma mansoni*. Vinte e cinco casos de esquistossomose cutânea ectópica foram encontrados descritos na literatura. A maioria dos pacientes era do sexo feminino, com idade média de 24,9 anos e predominância da raça branca. A localização mais comum foi o tórax anterior e abdômen. Freqüentemente, as lesões eram assintomáticas. Em alguns casos, no entanto, coexistiam alterações sistêmicas graves, relacionadas ao parasito, como mielite transversa e a forma aguda toxêmica da doença. Na maior parte dos casos, a infecção intestinal não pôde ser demonstrada. O reconhecimento destas lesões cutâneas pode ser importante por proporcionar oportunidade para um diagnostico etiológico nestes casos.

REFERENCES

1. BITTENCOURT, A.; PINHO, O.; LENZI, H. L. & COSTA, I. M. C. - Extragenital cutaneous lesions of Schistosomiasis mansoni. Report of two cases. **Amer. J. trop. Med. Hyg., 28**: 84-86, 1979.

2. CONVIT, J. & REYES, O. - Ectopic skin lesions produced by *Schistosoma mansoni*. **Amer. J. trop. Med. Hyg., 22**: 482-484, 1973.

3. COSTA, I. M. C.; MOREIRA, R. R. & MORAIS, M. A. P. - Esquistossomose cutânea ectópica. **An. bras. Derm., 64**: 183-184, 1989.

4. FINDLAY, F. H. & WHITING, D. A. - Disseminated and zosteriform cutaneous schistosomiasis. **Brit. J. Derm., 85 (suppl. 7)**: 98-101, 1971.

5. FURTADO, T.; VIEGAS, A. C. & CUNHA, A. S. - Esquistossomose cutânea ectópica por *Schistosoma mansoni*. In: REUNIÃO ANUAL DOS DERMATÓLOGOS LATINO-AMERICANOS, 5.. Buenos Aires, 1980. *apud* VALE, E. C. S. & FURTADO, T. - Esquistossomose cutânea ectópica por *Schistosoma mansoni*. **Med. cut. ibero lat.-amer., 15**: 233-237, 1987.

6. GUIMARÃES, N. S. & SOUZA, A. P. B - Esquistossomose cutânea ectópica. Três novos casos. **An. bras. Derm., 62**: 101-103, 1987.

7. GUIMARÃES, N. S.; SOUZA, A. P. B.; DINIZ, M. R. & GUIMARÃES, N. A. - Esquistossomose cutânea ectópica. **An. bras. Derm., 60 (supl. 1)**: 257-261, 1985.

8. JACYK, W. K.; LAWANDE, R. V. & TULPULE, S. S. - Unusual presentation of extragenital cutaneous schistosomiasis mansoni. **Brit. J. Derm., 103**: 205-208, 1980.

9. MACDONALD, D. M. & MORRISON, J. G. L. - Cutaneous ectopic schistosomiasis. **Brit. med. J., 2**: 619-620, 1976.

10. MILLIGAN, A. & BURNS, D. A. - Ectopic cutaneous schistosomiasis and schistosomal ocular inflammatory disease. **Brit. J. Derm., 119**: 793-798, 1988.

11. PATRUS, O. A. R. - **Esquistossomose cutânea ectópica**. Belo Horizonte, 1974. (Dissertação de Mestrado - Faculdade de Medicina da Universidade Federal de Minas Gerais).

12. PIVA, N.; CORREA, M. C. & DINIZ, V. - Lesões dermatológicas na esquistossomose mansoni. In: CONGRESSO DA SOCIEDADE BRASILEIRA DE MEDICINA TROPICAL, 14., João Pessoa, 1978. **Resumo**. p. 116.

13. TORRES, V. M. - Dermatologic manifestations of schistosomiasis mansoni. **Arch. Derm., 112**: 1539-1542, 1976.

14. VALE, E. C. S. & FURTADO, T. - Esquistossomose cutânea ectópica por *Schistosoma mansoni*. **Med. cut. ibero lat.-amer., 15**: 233-237, 1987.

15. WOOD, M. G.; SROLOVITZ, H. & SCHETMAN, D. - Schistosomiasis. Paraplegia and ectopic skin lesions as admission symptoms. **Arch. Derm., 112**: 690-695, 1976.

Received: 02 February 1998
Accepted: 11 May 1998