Caso Clínico

Ectopic cutaneous ***Schistosomiasis mansoni*** *in the State of Bahia, Northeast Brazil: report of 8 cases and literature review* *

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Resumo: Casos clínicos de esquistossomose cutânea ectópica, observados entre 1988 e 1996 são descritos em oito pacientes do Estado da Bahia, Brasil, perfazendo um total de 31 casos desta doença documentados em brasileiros. É feita uma breve revisão dos casos brasileiros anteriormente relatados.
Palavras-chave: Esquistossomose; pele; schistosoma mansoni.

Summary: Eight patients from the State of Bahia, Northeast of Brazil, with ectopic cutaneous schistosomiasis, seen between 1988 and 1996 are presented, amounting to a total number of thirty one cases of ectopic cutaneous schistosomiasis so far documented among Brazilians. A brief review of previously reported brazilian cases is presented.
Key words: Schistosomiasis; skin; schistosoma mansoni.

INTRODUCTION

The skin being exposed to the environment is often the portal of entry for parasites.[1] Parasitic infections with parasites characteristically can produce cutaneous lesions as well as systemic disease in humans. Schistosomiasis is the name given to a group of diseases caused by trematodes of the genus *Schistosoma*. Humans and other animals are definitive hosts, and snails of various genera are intermediate hosts.[2] The parasite penetrates through the skin where it produces erythema accompanied by pruritus, which is the first skin manifestation of the disease, hours after the cercarial penetration. Invasion is manifested by mild or sometimes fairly marked systemic disorders.[3] The *Schistosoma mansoni* larvae, after accessing the circulation, pass through the lungs, continue their "journey" to the left side of the heart, and finally reach the intrahepatic venous system where they mature into adult flukes. Then the worms migrate against the portal flow to the mesenteric and colonic venules where females are fertilized. They deposit their eggs in the mucosa and submucosa venules of the colon and rectum. During the stage of oviposition, the patients suffer generalized malaise, cough, fever, arthralgias, gastrointestinal disturbances, generalized lymphadenopathy and hepatosplenomegaly. In a low percentage of cases at this stage there may be urticaria, periorbital edema and purpuric lesions,[4] three to six weeks after the infection. The ova that accumulate in various organs, especially the colon, liver, spleen, and lungs incite a granulomatous reaction that eventually leads to fibrosis.[5,6] The major complication is portal hypertension provoked by obstructive portal lesions. It is only during the stage of granuloma formation that persistent cutaneous lesions, caused by deviations from the usual sites of egg deposition may occur. In endemic areas this probably happens within the first months of infection.[7] Papular, ulcerative, granulomatous and fistulous lesions may develop in the genital and perirectal skin in heavily infected individuals, secondary to the deposition of ova in dermal vessels contiguous to the pelvic vessels.[7] Only in rare instances the *S.mansoni* eggs are lodged in extragenital skin.[1,4,7,8,9] The disease is then called ectopic cutaneous schistosomiasis and

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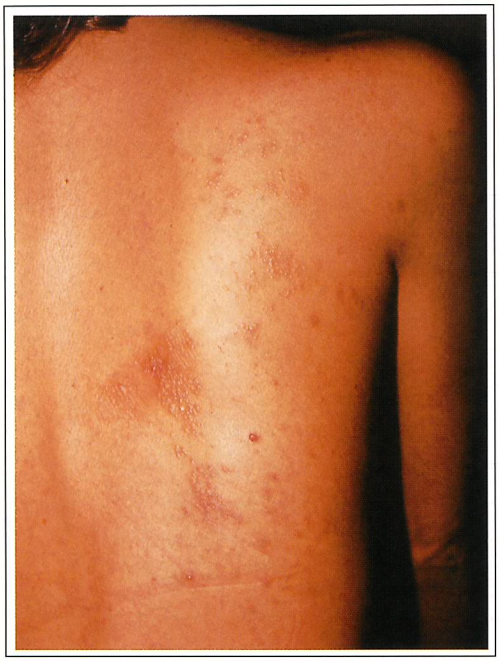

Figure 1: Multiple hyper-pigmented papules and plaques on the back (case 4)

is sometimes associated with neurological symptoms in cases of simultaneous cerebral and spinal cord involvement.[4,10] Interesting cases of spontaneous regression of the cutaneous lesions have been reported.[11]

Schistosomal cutaneous granulomas must be very rare indeed,[12] since there have been only a few cases reported in the literature, in spite of the fact that there are millions of people suffering from schistosomiasis. *Schistosomiasis mansoni* is widespread in Africa and Latin America. The State of Bahia, (Northeast Brazil) is highly endemic for this parasitosis.

In this article, eight new cases of cutaneous schistosomiasis are presented. The literature is reviewed, and the frequency, characteristics and physiopathology are discussed.

CASE REPORTS

Case 1

A 24-year-old white female, from Salvador, BA - Brazil, complaining of enlarging, pruritic eruptions on the trunk. All lesions had appeared within two or three days and had gradually enlarged in diameter and height over a three month-period. The patient denied having had body contact with potentially infested water. The lesions were firm, painless papules, with erythematous and lichenoid aspect, measuring about 5mm each, distributed on the left pectoral area, ascending to the left arm. Stool examination (4 examinations) were negative for *Schistosoma* ova. The white blood cell count was 4,400 per mm^3 with 12 percent eosinophils. Intradermal reaction for schistosomiasis test was strongly positive. Biopsy sections from a typical papule showed in the dermis multiple perivascular and periadnexial granulomas, centered by many viable *S.mansoni* eggs surrounded by a large eosinophilic necrotic area with peripheral macrophages, epithelioid and giant cells. Additionally lymphocytes and eosinophils were observed. The patient received 15mg/kg of oxamniquine in a single dose, followed by progressive regression of the lesions after the biopsy and treatment with some slight hyperpigmentation.

Case 2

A 27-year-old white female, from Salvador, BA - Brazil, presenting at consultation grouped papules in the left flank for three months after bathing in a potentially infective river. Stool examination revealed ova of *S.mansoni*. Biopsy showed schistosomal periovular granulomas within the dermis with necrotic areas. The patient received 15mg/kg of oxamniquine and showed progressive regression of the lesions.

Case 3

A 22-year-old black female, from Salvador, BA - Brazil, presenting erythematous and pruritic papules in the

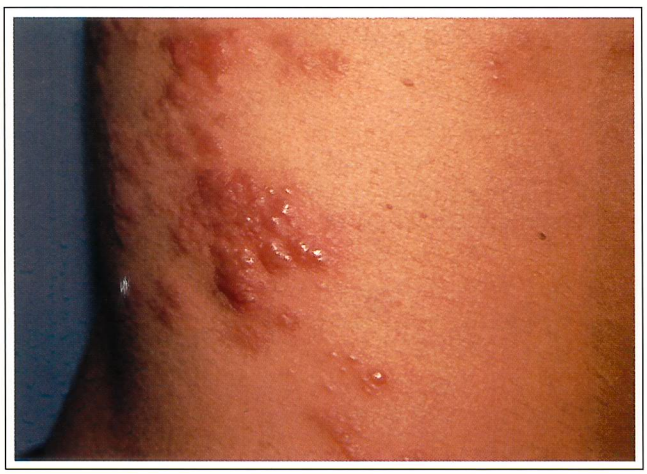

Figure 2: Typical distribution and appearance of the lesions in ectopic cutaneous schistosomiasis. Clusters of erythematous papules and plaques on the left pectoral region and back (case 5)

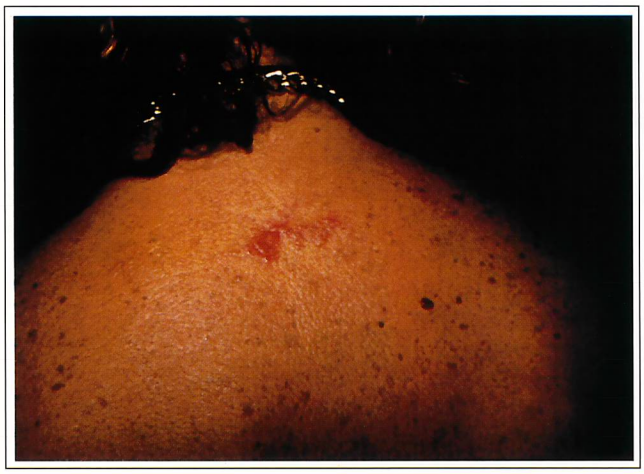

Figure 3: Grouped papules forming plaques on the back (case 6)

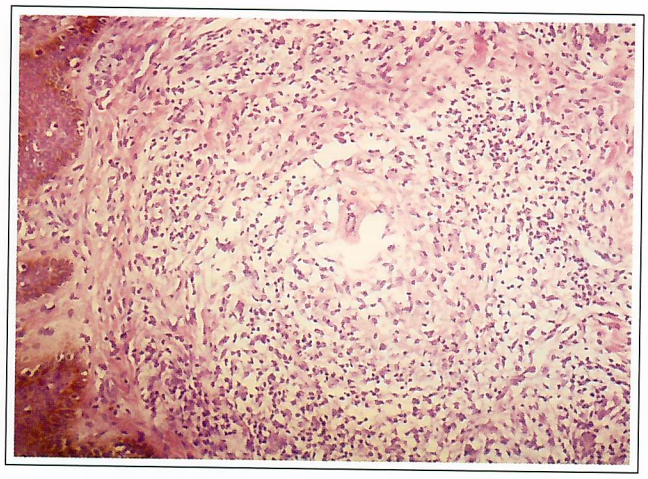

Figure 4: Skin biopsy: dermal schistosomal granuloma showing S. mansoni ovum in the center of a pallisading zone around central necrotic area. Histiocytes, lymphocytes and plasma cells were also seen (case 7 - HE, 250x)

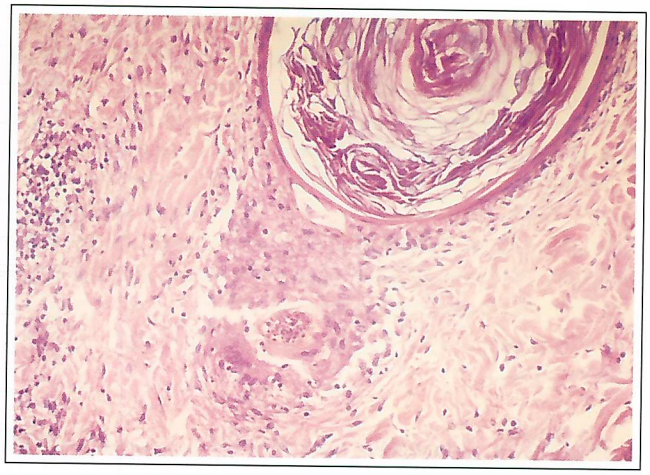

Figure 5: Isolated papule demonstrating schistosomal periovular granuloma in the dermis, with an infundibular cyst and an infiltrate of lymphocytes and eosinophils around them (case 7 - HE, 250x)

left pectoral region for ten days. The lesions appeared two months after bathing in potentially infectious water. Stool examination revealed ova of *S. mansoni*. Biopsy showed schistosomal periovular granulomas within the dermis with necrotic areas. The patient received 15mg/kg of oxamniquine with partial regression of the lesions.

Case 4

A 26-year-old white female, from Salvador, BA - Brazil, presenting during three months isolated and confluent papules forming right dorsal plaques, with a herpetiform and zosteriform aspect (Figure 1). Stool examinations (three examinations) were negative for *Schistosoma* ova. The intradermal reaction for schistosomiasis was positive (13mm). The histology of the lesions showed dermal granulomatous reaction around ova of *S. mansoni*. She was treated with 15mg/kg of oxamniquine in a single dose, resulting in progressive regression of the lesions.

Case 5

A 36-year-old white female, from Salvador, BA - Brazil, had for two months in her left pectoral region and back isolated or grouped erythematous papules, with a herpetiform aspect and with a zosteriform distribution (Figure 2). Pathological examination revealed dermal schistosomal granulomas. After treatment with a single dose of oxamniquine (15mg/kg) the lesions disappeared.

Case 6

A 45-year-old white female, from Salvador, BA - Brazil, presented for two months with central dorsal erythematous papules and plaques (Figure 3). This was unique in having lesions in the midline. More than one year before she had body contact with potentially infectious water in an endemic region. Stool examinations (five examinations) were negative for *Schistosoma* ova. Biopsy examination showed dermal periovular granulomas. The lesions cleared progressively after treatment with a single dose of oxamniquine.

Case 7

A 25-year-old white female, from Salvador, BA - Brazil, for two months complained of itchy, infiltrated erythematous papules on her left scapular region. One year before she had body contact with potentially infected water in an endemic region. Histology showed schistosomal periovular granulomas in the dermis, with eosinophilic necrotic areas, infundibular cyst and an infiltrate of lymphocytes and eosinophils around them (Figure 4, 5). The patient was treated with oxamniquine. The papules completely disappeared, leaving areas of post-inflammatory hyperpigmentation.

Case 8

A 12-year-old boy, from Feira de Santana, BA - Brazil, presented papules in his right posterior thigh for one

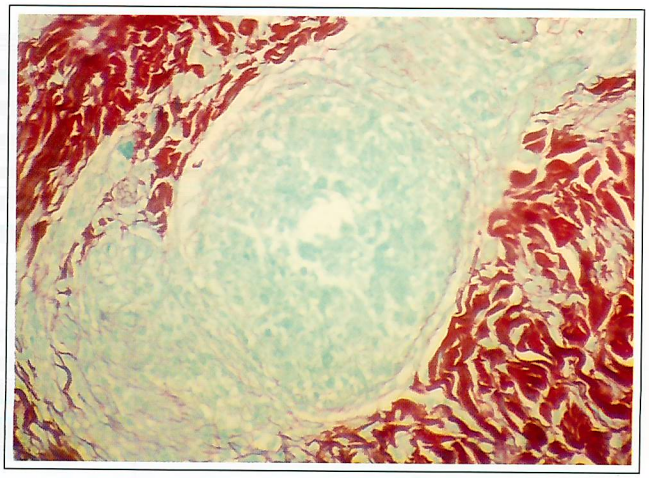

Figure 6: Papule showing scanty fibrous and fibrilar stroma at the periphery of one granuloma (case 8 - Sirius-red, 320x)

week. He had body contact with potentially infected water three months earlier. Pathological examination revealed dermal granulomas around large necrotic areas, centered around *Schistosoma* ova. The granulomas were densely packed collections of cells. Macrophages and eosinophils were the predominant cell-types, but plasma cells and lymphocytes were also observed. The fibrous and fibrilar stroma was scanty and tended to be more concentrated at the periphery of the granulomas (Figure 6). An infundibular cyst was present around a granuloma. The lesions regressed progressively after treatment with a single dose of oxamniquine.

DISCUSSION

Prior to the present report, in the past 25 years only 23 cases of ectopic cutaneous schistosomiasis have been reported in Brazil (Table 1). The eight cases presented here bring to 31 the cases of ectopic cutaneous schistosomiasis so far documented among Brazilians. Among all these cases, 19 were from the State of Bahia, six from Minas Gerais, four from Pernambuco and two from Sergipe. Most of them had a recent history of body contact with potentially infested water. In our cases, it may be noted that all of them had their residences in urban areas, without the nearby presence of known infested water sources. The overall male to female

Table 1: Features of 31 Brazilian patients with ectopic cutaneous schistosomiasis

Author	Year	State	Age	Sex	Probable time since contact with infested water (months)	Clinical presentation
Patrus[10]	1974	MG	04	F	6	Spinal schistosomiasis. Papules, right foot
			26	F	4	Papules, right hemithorax & abdomen, zosteriform
			15	F	3	Papules, right hemithorax, zosteriform distribution
Piva et al.[16]	1978	SE	32	M	?	Nodule, posterior chest
			17	F	> 12	Pregnant 4th month. Cutaneous rush. Plaques and papules, anterior and posterior chest
Bittencourt et al.[20]	1979	BA	26	F	1	Erythematous papules, lumbar region
			28	F	> 12	Confluent papules, back
Andrade[21]	1979	BA	15	F	?	Plaque, left side of the nose
Furtado et al.[22]	1980	MG	?	?	?	Papules, thorax, zosteriform distribution
Guimarães et al.[23]	1985	BA	22	F	8	Papules, sternocleidomastoid region
			33	F	?	Papules, left lateral abdomen
			23	M	8	Papules, left lateral abdomen, buttocks and scrotum
			30	F	> 12	Papules, left mammary region and abdomen
Vale & Furtado[24]	1987	MG	29	F	1	Papules, periumbilical region
Guimarães & Souza[11]	1987	BA	29	F	3	Papules, left pectoral region, zosteriform distribution
			32	F	?	Confluent papules, right pectoral region, herpetiform aspect
			23	F	1	Papules, right pectoral region, zosteriform distribution
Barata et al.[25]	1988	MG	39	F	> 12	Papules in mamoplasty scar
Costa et al.[26]	1989	BA	28	M	3	Papules, right side of malar region
Lima et al.[27]	1995	PE	20	M	?	Papules, left lumbar region
			32	F	?	Papules, left flank
			18	M	> 12	Papules and plaques, right flank, zosteriform
			41	F	> 12	Papules, left abdominal region, zosteriform
Case 1	1997	BA	24	F	?	Papules, left pectoral region and left arm
Case 2			27	F	3	Papules, left flank
Case 3			22	F	2	Papules, left pectoral region
Case 4			26	F	3	Isolated papules and plaques, right lumbar region
Case 5			36	F	2	Grouped papules, left pectoral region and back,
Case 6			45	F	> 12	Papules, central region of the back
Case 7			25	F	12	Papules, left scapular region
Case 8			12	M	3	Papules, right posterior region of thigh

ratio was approximately 1:4, with a mean age of 26±9 (SD) years.

The histological pattern of our patients was similar between the cases and did not differ from the previous related Brazilian cases. Skin periovular granulomas present peculiar histological features. They are represented by a cluster of cells with very scanty extracellular matrix at the periphery of the granulomas. They are smaller than liver granulomas, with less fibrosis. Differences between the periovular skin granulomas and the granulomas of other sites could be expected in view of the diverse cellular and organ specific composition. A similar situation was observed in experimental intestinal schistosomiasis[13] and in experimental pulmonary schistosomiasis[14] in sharp contrast to the hepatic periovular granulomas. Such differences concerning the formation of fibrosis in schistosomal periovular granulomas could be related to the presence of fat-storing cells in the liver. These cells are differentiated in myofibroblasts and fibroblasts and they are a constituent of granulomas.[15] It is remarkable that in the related Brazilian cases the skin schistosomal periovular granulomas had frequently extensive necrosis. This could indicate that most of these granulomas were in the early necrotic-exudative phase at the time of diagnosis. It is curious that in some cases the granulomas surround infundibular cysts, as demonstrated in at least two previously reported cases[10,16] and in two of our patients (Case 7, 8). These could occur as a result of mechanical occlusion of the follicular orifice by the dermal periovular granulomas, associated with inflammation, resulting in progressive cystic ectasia of the hair follicle infundibulum.

S. mansoni adults live in the mesenteric venous system or intrahepatic portal vessels and less frequently in the hemorrhoidal plexus. Unusual ectopic lesions due to migration of schistosoma worms and oviposition can occur anywhere in the body.[7] Various explanations have been proposed for the ectopic location of eggs and worms in schistosomiasis. It has been postulated that they reach the skin via anastomosis by pelvic plexuses of veins, porto-caval anastomosis, the vertebral venous system or pulmonary arteriovenous anastomosis. Chronic and massive infections may cause arteriovenous fistulas, which permit eggs to pass from the pulmonary to the systemic circulation, accessing the small skin arteries. Congenital heart defects and congenital or acquired vascular anastomosis also can produce such a pattern.[4,7,8,9,17,18] A transient connection between the vertebral venous system with the vesical and hemorrhoidal veins may occur during elevation of thoracoabdominal pressure consequent to heavy coughing or forceful defecation[8,12] or the application of a constricting device around the abdomen.[4] It would explain the simultaneous appearance of multiple lesions.

Asymptomatic persistent extragenital cutaneous granulomatous lesions present as papules or plaques, arranged in groups at times with a herpetiform aspect or in a zosteriform distribution, usually on the shoulders, pectoral region, neck, face and umbilicus.[7] In all our female patients, ectopic cutaneous lesions affected the trunk, laterally situated in the great majority. The exception was the oldest of these patients, who presented medial lesions. In our study, the male to female ratio was 1:7. Due to the small number of cases and the lack of data on the level and frequency of exposure it is not possible with certainty to draw any conclusion concerning the prevalence of infection between males and females. In spite of this, it may be speculated that there is some special aspect of the female anatomical vasculature that may facilitate an ectopic distribution of the ova and could perhaps explain this difference.

It has been suggested that ectopic forms of the disease are not easily diagnosed on clinical grounds without histological examination[18,19] and because of this fact probably many cases are not diagnosed and consequently not reported. However, in seven of our cases, the clinician initially diagnosed schistosomiasis prior to finding the ova of *Schistosoma mansoni* in histological sections of the skin lesions. None of our patients had the severe form of schistosomiasis or manifested neurological symptoms. They have been successfully treated with antischistosomal drugs.

These infrequent but quite typical lesions allow an early diagnosis and treatment of the disease if cutaneous schistosomiasis is suspected by clinicians, when individuals in or from endemic areas have persistent papulonodular asymmetrical lesions on the perineum or trunk. ❑

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