Revista da Sociedade Brasileira de Medicina Tropical 41(6):668-671, nov-dez, 2008

# Ectopic schistosomiasis: description of five cases involving skin, one ovarian case and one adrenal case

## Esquistossomose ectópica: descrição de cinco casos de envolvimento da pele, um do ovário e um da glândula supra-renal

**Wendell Luiz Santos Poderoso[1], Wagner Barreto de Santana[1], Emerson Ferreira da Costa[2], Rosana Cipolotti[2] and Ricardo Fakhouri[2]**

**ABSTRACT**

Seven cases of patients with ectopic schistosomiasis from the State of Sergipe, Brazil, are presented (five involving skin, one ovarian and one adrenal). Data were collected from surveying the clinical records and anatomopathological reports in the files of the dermatology and pathology clinics of the University Hospital of the Federal University of Sergipe, from 1995 to 2005. The patients' mean age at diagnosis was 21.1 years. In the dermatological cases, full cures were achieved after treatment with oxamniquine. In the ovarian case, there was an association with embryonic carcinoma: this patient underwent surgery with adjuvant chemotherapy and praziquantel treatment, with satisfactory evolution. The adrenal case was associated with adenoma.

***Key-words:*** Ectopic schistosomiasis. *Schistosoma mansoni*. Ovary. Adrenal. Skin.

**RESUMO**

São apresentados sete casos de esquistossomose ectópica (cinco de pele, um de ovário e um de supra-renal) procedentes do Estado de Sergipe, coletados a partir de pesquisa de prontuários e laudos anátomo-patológicos nos arquivos dos Serviços de Dermatologia e de Patologia do Hospital Universitário da Universidade Federal de Sergipe, entre os anos de 1995 e 2005. A média de idade dos pacientes ao diagnóstico foi de 21,1 anos. Nos casos dermatológicos, houve melhora total das lesões após tratamento com oxamniquine. No caso de ovário houve associação com carcinoma embrionário; a paciente foi submetida à cirurgia com quimioterapia adjuvante e praziquantel, evoluindo satisfatoriamente. O caso de supra-renal estava associado a adenoma.

***Palavras-chaves:*** Esquistossomose ectópica. *Schistosoma mansoni*. Ovário. Supra-renal. Pele.

Although *Schistosoma mansoni* is the single species of trematode causing schistosomiasis in Brazil, more than 30 million individuals are exposed to the disease, and around 3 to 4 million may be infected[1,2,11].

Ectopic schistosomiasis presentations occur when the parasite eggs or adult forms are located far from their normal site (the portal system). Ectopic schistosomiasis has been reported in the cecal appendix, gallbladder, pancreas, peritoneum, urogenital system, central nervous system, myocardium, skin, esophagus, stomach, thyroid and adrenal[1,4].

Central nervous system involvement occurs both with and without symptoms[15]. Following high infestation rates, major lung involvement with respiratory symptoms is usually reported[17]. The urogenital presentation is not uncommon in endemic areas, and may be associated with local cancers[4]. The skin may be involved, but is not associated with malignant tumors[10].

We report seven cases of ectopic schistosomiasis from the databases of the dermatology and pathology clinics of the University Hospital of the Federal University of Sergipe, between 1995 and 2005. The clinical findings and pathological characteristics are described, with emphasis on the organ or tissue involved.

## CASE REPORTS

The general data on the cases, such as gender, age, place of professional activity, body affected and diagnosis are presented in Table 1.

**Cases involving skin.** There were five cases of cutaneous schistosomiasis. The patients' ages ranged from 11 to 36 years. In four cases, the anterior part of the thorax was involved and the lesion had disseminated to the neck and face (case 1), scapula

1. Curso de Medicina, Universidade Federal de Sergipe, Aracaju, SE. 2. Hospital Universitário, Universidade Federal de Sergipe, Aracaju, SE.
***Address to:*** Dr. Wendell Luiz Santos Poderoso. Rua Estância 1382, Bairro Cirurgia, 49055-000 Aracaju, SE.
Tel: 55 79 3213-0733
e-mail: wendellpoderoso@gmail.com
Recebido para publicação em 08/04/2008
Aceito em 29/10/2008

*Table 1 - General data on cases: gender, age, location, professional activity, body part affected and diagnostic tool.*

| Case | Gender | Age (years) | Origin | Professional activity | Organ | Diagnostic tool |
|---|---|---|---|---|---|---|
| **1** | male | 19 | Aracaju | typist | skin | incisional biopsy |
| **2** | female | 17 | Aracaju | student | skin | incisional biopsy |
| **3** | male | 28 | Aracaju | driver | skin | incisional biopsy |
| **4** | male | 11 | Aracaju | student | skin | incisional biopsy |
| **5** | male | 36 | Umbaúba | farmer | skin (perianal) | incisional biopsy |
| **6** | female | 15 | Umbaúba | fisherman | ovary | anatomical specimen |
| **7** | male | 22 | Estância | student | adrenal | anatomical specimen |

(case 2) and left inguinal region (case 4). In case 3, the skin lesions followed the metamers and were concentrated in the left hemithorax. In case 5, the perianal region was the only site involved.

Four cases presented as papular lesions (cases 1 to 4), and in case 1, with acneiform skin color presentation. Two cases had vesicular lesions (cases 2 and 3). In case 2, the lesions tended to coalescence, in plate shape. Only case 4 showed uniform papular lesions (Figure 1). Case 5 had an expansive lesion that projected into rectum, going beyond the orificium fistulae in the perianal region.

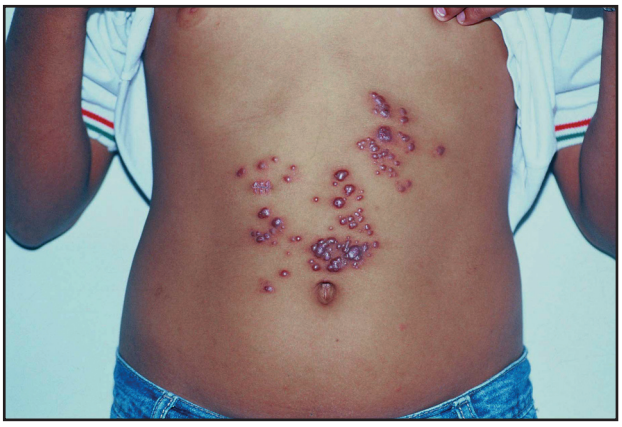

*Figure 1 - Papules in epigastric region.*

Pain was the most frequent complaint (cases 3 to 5), associated with local stinging and pruritus in cases 3 and 4. Fever was reported only in the case with anal involvement. Case 1 did not report pruritus, but mentioned nausea, headache, vomiting and sporadic diarrhea. Cases 3 and 4 did not report any systemic complaints.

Pathological examinations showed schistosomotic granuloma in all cases (Figure 2). Case 2 presented granuloma around *Schistosoma mansoni* eggs, surrounded by an extensive necrotic area composed of eosinophil. Case 5 had calcified eggs and case 3 only had eggshells. Cases 2 and 4 had predominant eosinophilic infiltrate, suggesting acute phase disease.

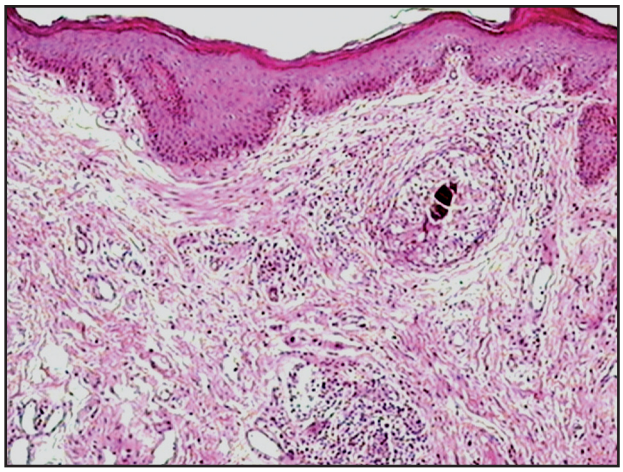

*Figure 2 - Micrograph showing schistosomotic granuloma.* **HE 400X.**

**Ovarian case.** One adolescent sought medical care because of hypogastric pain, ascites and unspecific stomach symptoms. She also reported weakness and weight loss. Abdominal echography identified a tumor involving the right ovary, Fallopian tube and epiploon. The patient underwent tumorectomy. Pathological examination of the ovary showed fibrous and enlarged tunica albuginea. A tumor was observed in the cortex, composed of primitive cells with enlarged nuclei, prominent multiple nucleoli, poor in cytoplasm, high mitotic index and vascular invasion. The cells were distributed in small nests or abortive glandular structures. The fallopian tube and epiploon were compromised by tumor cells, and viable *Schistosoma mansoni* eggs were found in the epiploon and right ovary.

**Adrenal case.** This patient was admitted for medical evaluation because of weakness, diffuse paresthesia, visual disturbances and hypertensive crisis. Laboratory tests showed hypokalemia and low serum renin level. Abdominal echography and computed tomography showed a tumor in the right adrenal gland (Figure 3). After tumorectomy (Figure 4), pathological examination identified tumor cells in the adrenal cortex, showing mixed, trabecular and acinar growth patterns. They showed regular morphology, with small, lightly hyperchromatic nuclei, and enlarged, light and thinly vacuolated cytoplasm. Mitosis

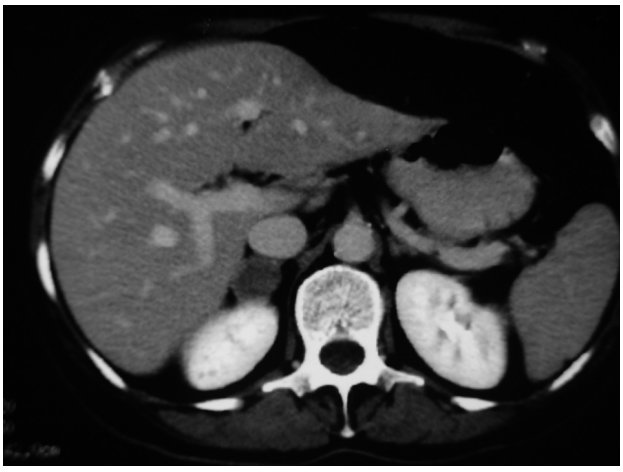

*Figure 3 - Axial computed tomography image showing adrenal mass.*

figures were rare, and some granulomas with viable *Schistosoma mansoni* were found (Figure 5). The final diagnosis was right adrenal cortex adenoma plus adrenal ectopic schistosomiasis.

After diagnosis, all patients received oxamniquine and remained asymptomatic during the clinical follow-up.

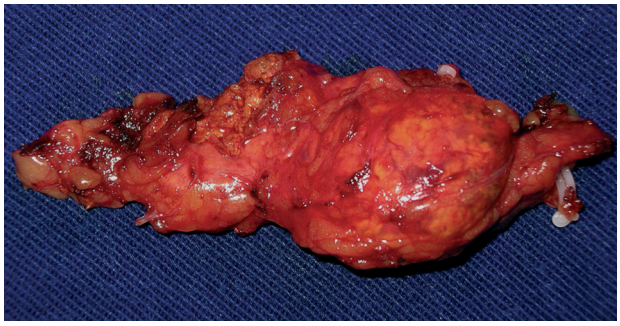

***Figure 4 - Macroscopic adrenal appearance after surgery.***

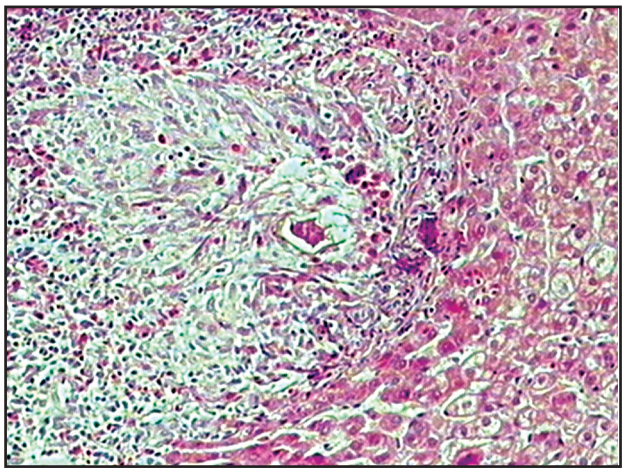

***Figure 5 - Micrograph showing tumor cells at adrenal cortex and Schistosoma.***

## DISCUSSION

We have presented personal data (age, gender, origin and professional activity), diagnoses and organs involved in seven cases of ectopic schistosomiasis (Table 1).

All the patients were 20 to 30 years old, which was in line with data in the literature. Although Fitzpatrick & Hook[9 18] described ectopic cutaneous schistosomiasis mainly among children, only one of our patients was a child, of eleven years of age.

Andrade-Filho[2] observed female predominance among ectopic cutaneous schistosomiasis patients (16 out of 25 cases). In Sergipe, the incidence was higher among men[19] and in our sample, five out of the seven patients were men. We attribute these findings to the male behavioral profile, which predisposes towards parasite exposition.

Among our five patients with ectopic cutaneous schistosomiasis, one had an anal lesion and the other four had thoracic involvement, one of them also with a frontal lesion. This location had been described by Fitzpatrick previously[8]. The patients' complaints varied, ranging from asymptomatic to painful lesions. This variability of presentations and symptoms had not been observed previously: slight pruritus had been the main reported complaint[10]. This study reported initial lesions as papules, progressing to nodules[10]. Our patients also presented papular-vesicular and acneiform lesions. For all five cases, pathological examination was essential for concluding the diagnosis. All the cases were treated with oxamniquine and were considered cured, with only hypochromic macula as residual features.

None of the patients presented any kind of complications, like secondary infections, ulcers or fistulas. Cases 2 and 4 seemed to be in the acute phase of the disease because, although they did not present acute disease signs or symptoms, pathological examinations showed granulomas with *Schistosoma mansoni* eggs inside, covered by a large necrotic area with eosinophils. In addition, case 4 reported contact with contaminated river water 15 days before the symptoms started[12].

Among the three *Schistosoma* species infecting man, only *Schistosoma mansoni* exists in the Americas[16]. Ramos[17] reported individuals carrying parasites or their eggs in their skin, but without exhibiting clinical symptoms. Therefore, although Sergipe is an endemic region for schistosomiasis, the frequency of diagnosed cases of ectopic cutaneous lesions is very low. *Schistosoma haematobium* is the species that most frequently lays eggs on the skin[17].

The cases with adrenal and ovarian findings were diagnosed incidentally. Both cases had these organs surgically removed because of their associations with neoplastic lesions.

Adrenal schistosomiasis was detected by means of pathological examination for adenoma. We did not find any previously reported cases of adrenal ectopic schistosomiasis. There are some reports showing female genital lesions caused by *Schistosoma mansoni,* mostly published between 1940 and 1960[3 5].

In a systematic review, Feldmeier[7] found 83 reports of women with genital involvement. The study revealed that the ovary was the organ most commonly affected, followed by the cervix, uterine tubes and uterus. Vulvar lesions corresponded to 6% of the cases reported. Nevertheless, Feldmeir[7] speculated that this proportion of vulvar cases did not represent the real female genital involvement rates. In his opinion, this reflected underestimation of internal genital disease.

Schistosomiasis may be associated to other ovarian diseases. Lee[13] reported the case of a patient with endometriosis and hemorrhagic lesions associated with ovarian ectopic schistosomiasis caused by *Schistosoma japonicum*. Eggs may be found inside the uterine tubes and uterus as well, and such findings may be important during pregnancy, when myometrial lesions, menorrhagia, spontaneous abortion and preterm delivery may result[6].

Schistosomotic parasites associated with cancer have been described previously (prostatic adenocarcinoma, blade squamous cell carcinoma or colorectal cancer)[8]. It was recently reported that, when schistosomiasis is found in the cervical channel with or without papillomavirus associated, progression to cervical cancer

may occur[8]. Case 6 showed ovarian cancer, which is histologically an embryonic carcinoma, in association with schistosomiasis. Swai[20] reported two cases in which *Schistosoma haematobium* was associated with ovarian cancer, without mentioning the histological profile. We could not find any reports on associations between ovarian tumors and *Schistosoma mansoni*.

Cases 5 and 7, were respectively a farmer and a fisherman who were living in rural areas and were accustomed to continuous exposure to a parasite-contaminated environment due their professional activities. The other patients were from an urban area, and they became contaminated during recreational activities.

The cases presented here indicate that ectopic presentations of *Schistosoma mansoni* infection may occur during chance contacts, without the chronic exposure that is usually associated with professional activities. In endemic regions, ectopic presentations must be taken into consideration and investigated in all compatible cases.

**ACKNOWLEDGEMENTS**

We thank Dr. Antônio Alves Júnior for the pictures of the adrenal schistosomiasis case.

**REFERENCES**

1. **Amaral RS, Porto MAS. Evolução e situação atual do controle da esquistossomose** no Brasil. **Revista da Sociedade Brasileira de Medicina Tropical 27:73-90,** 1994.
2. **Andrade Filho S, Lopes MS, Corgozinho Filho AA, Pena GP. Ectopic cutaneous** schistosomiasis: report of two cases and a review of the literature. Revista do Instituto de Medicina Tropical de São Paulo 40:253-257, 1998.
3. Arean VM. Manson's schistosomiasis of the female genital tract. American Journal of Obstetrics and Gynecology 72: 1038-1056, 1956.
4. **Basílio-de-Oliveira CA, Aquino A, Simon EF, Eyer-Silva WA. Concomitant** prostatic schistosomiasis and adenocarcinoma: case report and review. The Brazilian Journal of Infectious Diseases 6:45-49, 2002.
5. **Câmara A. Esquistossomose genital feminina. Revista de Ginecologia e** D'Obstetrícia 3: 271-280, 1959.
6. Edington GM, Nwabuebo I, Junaid TA. The pathology of schistosomiasis in Ibadan, Nigeria with special reference to the appendix, brain, pancreas and genital organs. Transactions of the Royal Society of Tropical Medicine and Hygiene 69:153-156, 1975.
7. Feldmeier H, Daccal RC, Martins MJ, Soares V, Martins R. Genital manifestations of schistosomiasis mansoni in women: important but negleted. Memórias do Instituto Oswaldo Cruz 93: 127-133, 1998.
8. Feldmeier H, Krantz I, Helling-Giese G, Rohrbach C. Preventing cervical cancer - a global issue. Nature Medicine 2: 1054, 1996.
9. Fitzpatrick TB. Dermatology in General Medicine. McGraw-Hill, New York, 1999.
10. Guimarães NS, Barbosa AAJ. Ectopic Cutaneous Schistosomiatosis Mansoni in the State of Bahia: Northeast Brazil: report of 8 cases and literature review. Anais Brasileiro de Dermatologia 72:375-380, 1997.
11. **Katz N, Peixoto SV. Análise crítica da estimativa do número de portadores** de esquistossomose mansoni no Brasil. **Revista da Sociedade Brasileira de** Medicina Tropical 33:303-308, 2000.
12. **Lambertucci JR, Rayes AAM, Barata CH, Teixeira R, Gerspacher-Lara R.** Acute shistosomiasis: report on five singular cases. Memórias do Instituto Oswaldo Cruz 92: 631-635, 1997.
13. Lee KF, Hsueh S, Tang MH. Schistosomiasis of the ovary with endometriosis and corpus hemorrhagicum: a case report. Chang Gung Medical Journal 23:438-441, 2000.
14. **Melo AL, Coelho PMZ.** *Schistosoma mansoni. In*: Neves DP, Melo AL, Genaro O, Linardi PM (eds) Parasitologia Humana, 10ª edição, Editora Atheneu, São Paulo, p. 174-193, 2000.
15. **Moreno-Carvalho OA, Nascimento-Carvalho CM, Bacelar AL, Andrade-Filho A,** Costa G, Fontes JB, Assis T. Clinical and cerebrospinal fluid (CSF) profile and CSF criteria for the diagnosis of spinal cord schistosomiasis. **Arquivos de Neuro-**Psiquiatria 61:353-358, 2003.
16. **Prata A. Esquistossomose Mansoni.** *In*: Veronesi R, Focaccia R (eds) Tratado de Infectologia, 2ª edição, Editora Atheneu, São Paulo, p.1374-1403, 2004.
17. **Ramos SF. Bilharzíose cutânea tardia.** Anais do Instituto de Higiene e Medicina Tropical 1:1-4, 1973.
18. Rook A. Textbook of Dermatology. Blackwell Scientific Publications, London, 1986.
19. **Secretaria de** Vigilância em Saúde. Programa de Controle da Esquistossomose [dados na Internet]. SVS [acesso 2007 nov. 13]. Disponível em: http://portal.saude.gov.br/portal/arquivos/pdf/casosesquistossomose.pdf, Ministério da Saúde, Brasília, 2007.
20. Swai B, Poggensee G, Mtweve S, Krantz I. Female genital schistosomiasis as an evidence of a neglected cause for reproductive ill-health: a retrospective histopathological studyfrom Tanzania. BioMedCentral Infectious Diseases 6:134, 2006.