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

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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.


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 à adenoma.


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 infected¹²¹¹. 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 adrenal¹⁴. Central nervous system involvement occurs both with and without symptoms¹⁵. Following high infestation rates, major lung involvement with respiratory symptoms is usually reported¹⁷. The urogenital presentation is not uncommon in endemic areas, and may be associated with local cancers⁴. The skin may be involved, but is not associated with malignant tumors¹⁰.

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

<table>
<thead>
<tr>
<th>Case</th>
<th>Gender</th>
<th>Age (years)</th>
<th>Origin</th>
<th>Professional activity</th>
<th>Organ</th>
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<tr>
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<tr>
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<td>22</td>
<td>Estância</td>
<td>student</td>
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</tr>
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</table>

(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.

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.

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
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.

### 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 & Hook² described ectopic cutaneous schistosomiasis mainly among children, only one of our patients was a child, of eleven years of age.

Andrade-Filho² observed female predominance among ectopic cutaneous schistosomiasis patients (16 out of 25 cases). In Sergipe, the incidence was higher among men³⁸ 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 previously⁹. 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 complaint¹⁰. This study reported initial lesions as papules, progressing to nodules¹⁰. 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 started¹².

Among the three *Schistosoma* species infecting man, only *Schistosoma mansoni* exists in the Americas¹⁶. Ramos¹⁷ 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 skin¹¹.

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 1960¹⁵.

In a systematic review, Feldmeier⁷ 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, Feldmeier 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. Lee¹³ 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 result⁶.

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. Case 6 showed ovarian cancer, which is histologically an embryonic carcinoma, in association with schistosomiasis. Swai 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**

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**REFERENCES**


