

Schistosomiasis mansoni: associated hepatosplenic, cardiovascular, and chronic miliary forms

Esquistossomose mansoni: formas hepatoesplênica, cardiovascular e miliar crônica associadas

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ABSTRACT

Introduction: The authors describe the main histopathological findings in a serious case of schistosomiasis. **Methods:** the study was based on the histological analysis of fragments from organs of a 26-year-old patient. **Results:** the necropsy revealed the association of forms: hepatosplenic, cardiovascular, and chronic miliary with eggs found in the glomerulus afferent arteriole and the rare association of locations in the myocardium, kidney, and pancreas.

Key words: Schistosomiasis; Schistosomiasis mansoni, Schistosoma mansoni; Liver Diseases; Heart Diseases.

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RESUMO

Introdução: os autores descrevem os principais achados anatomopatológicos em um caso grave de esquistossomose. *Métodos:* o estudo baseou-se na análise histológica de fragmentos de órgãos de paciente de 26 anos de idade. *Resultados:* a necropsia revelou associação de formas: hepatoesplênica, cardiovascular e miliar crônica, com encontro de ovos na arteríola aferente do glomérulo e a rara associação de localizações no miocárdio, rim e pâncreas.

Palavras-chave: Esquistossomose; Esquistossomose mansoni; Schistosoma mansoni; Hepatopatias; Cardiopatias.

INTRODUCTION

Despite the absence of clinical data, which prevents an anatomoclinical correlation analysis, this case is worthy of record due to its singularity observed at necropsy.

CASE REPORT

This is patient with hepatosplenic schistosomiasis mansoni, of Symmers-Bogliolo, associated with the cardiopulmonary form, accompanied by the dissemination of eggs in all examined organs (miliary chronic dissemination) particularly intense in the lungs, pancreas, and large intestine. The existence of a couple of worms was observed in the lumen of the vein in the peripancreatic fat tissue, indicating likely oviposition in the pancreas of one egg in the lumen of an afferent arteriole of the glomerulus and myocardium granulomas.

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The major alterations were:

- liver: chronic granulomatous schistosomiasis inflammation (granuloma in the exudative phase) and moderate fibrosis in portobiliary spaces, without invading or subverting the lobular architecture (Figure 1). Lesion caused by a dead worm, showing traces of the worm surrounded by extensive necrosis and increased inflammatory exudate.

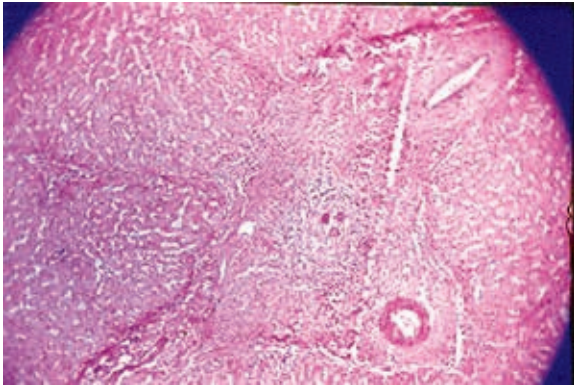


Figure 1 - Fibrosis and productive granulomas in the porta space.

- lung: numerous granulomas, sometimes confluent, small, at the stage of fibrosis, involving or not eggs or their shells in the lumen, on the wall, or in the adventitious of small arteries or arterioles; associated with reduction or occlusion of veins' lumen. Wall necrosis in some arterioles (Figure 2);

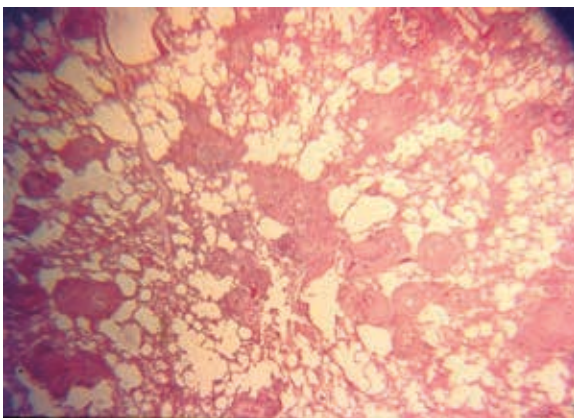


Figure 2 - Lung: numerous productive granulomas and in healing through fibrosis.

- heart: chronic granulomatous myocarditis (granuloma in the exudative productive phase centered by an eggshell partially surrounded by giant cell of foreign body type and more externally by epithelioid macrophages and eosinophils); and

eosinophilic exudate, discreet, between heart fiber cells, which were generally well preserved. Rare cells in necrosis, isolated, in the vicinity of the granuloma (Figure 3);

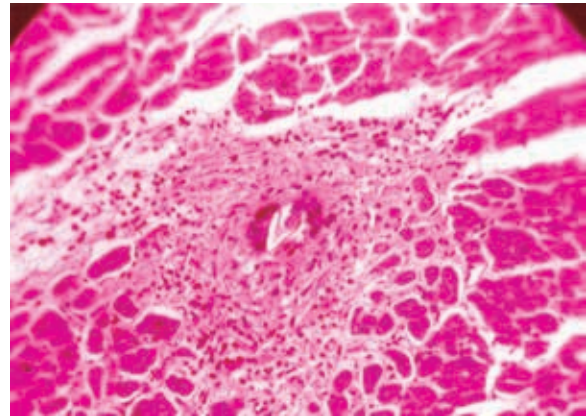


Figure 3 - Granuloma in the productive phase in the myocardium.

- pancreas: a pair of adult worms in the lumen of a vein in the peripancreatic fat, without inflammatory reaction around the vase. Several eggs wrapped in chronic inflammatory reaction without forming typical granulomas in the parenchyma. The inflammatory exudate infiltrated and destroyed part of the lobes and groups of cells, with a partial disorganization of lobular architecture. Interstitial and moderate intralobular fibrosis; perineuritis and discrete ductile hyperplasia (Figure 4);

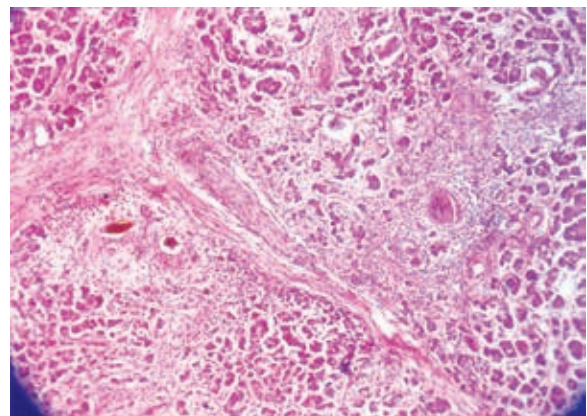


Figure 4 - Pancreas: granulomas, anti-inflammatory infiltrate, fibrosis and destruction of part of the parenchyma.

- kidney: eggshell in the lumen of an afferent arteriole in a glomerulus (Figure 5), thickening of the basal membrane and proliferation of mesangial

cells; discreet infiltrate of mononuclear cells in the vicinity of the arteriole; granuloma in the exudative phase around a deformed egg in the interstice (Figure 6); and mild interstitial subcapsular fibrosis. Tubules without notable alterations.

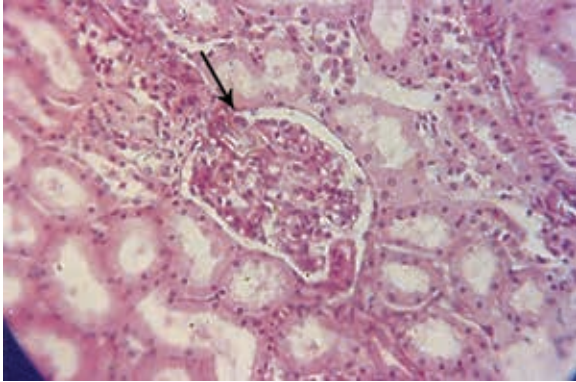


Figure 5 - *S. mansoni* egg in the lumen of an afferent arteriola (arrow).

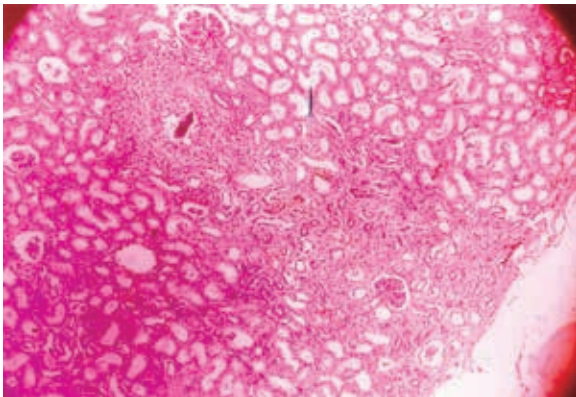


Figure 6 - Kidney: granuloma in the interstice.

- large intestine: large number of eggs (about 30 per microscopic field) in the submucosa and few in the mucosa lamina propria, largely calcified, without inducing the characteristic granulomatous reaction. Nonspecific inflammatory reaction, diffuse, with a predominance of eosinophils and mononuclear cells, more intense at the level of the mucosa and denudation of the coating epithelium (Figure 7).

DISCUSSION

A case of severe schistosomiasis mansoni with association of forms is presented - hepatosplenic, in the lungs, heart, intestines, kidney, and pancreas - in

26-year-old man necropsied at the Santa Casa de Misericórdia de Belo Horizonte, Minas Gerais.

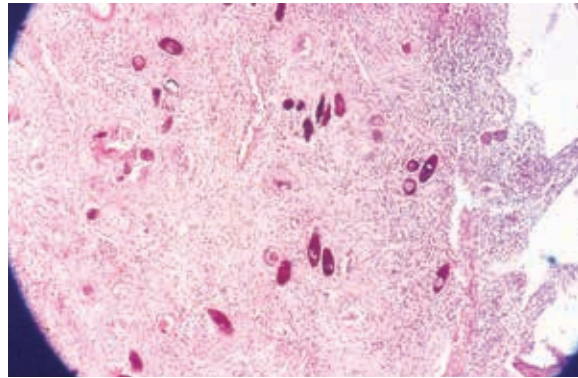


Figure 7 - Large intestine: numerous calcified eggs in the submucosa.

The unusual of these alterations consists in the anatomopathological alterations and unusual location of eggs and worms, some of them never reported.

Ectopic lesions are considered those caused by worms or eggs outside the portomesenteric system (hepato-intestinal system), habitat of adult *Schistosoma mansoni* in the human body.¹⁻¹² Faust⁷, however, excluded pulmonary arterioles lesions from ectopic sites. The frequency of pulmonary locations varies between the different regions of origin of patients.^{5,6,9,10-20} It is assumed that 20% of schistosomal patients have pulmonary lesions, however, only a small percentage of these develops cor pulmonale.^{5,6,9-20}

The fundamental anatomic substrate responsible for hemodynamic alterations in the hypertension pulmonary circulation and cor pulmonale forms are vascular alterations in the level of small arterioles and arteries. Thus occurs in a small number of cases and is followed, almost invariably, by the hepatosplenic form as the result of embolization of eggs and worms in the pulmonary artery resulting from the portosystemic circulation.^{6,20} It is assumed that eggs coming from the portal vein arrive at the lungs through the collaterals that form between the porta system and systemic circulation in order to compensate for the increased portal pressure.^{8,18} This has been confirmed experimentally.^{3,4} It is understandable, therefore, that serious pulmonary lesions may occur, as in fact occur, during the development of the hepatosplenic form through the establishment of communication between the portal and systemic circulation (hepatoscaping routes) that promote direct and continuous drainage of eggs to the right atrium and, consequent-

ly, the constant penetration of eggs in the pulmonary tree. The number of eggs found in the lungs is disproportionate to the number of worms, whose finding is infrequent, indicating that in general postures occur outside the pulmonary tree.^{21,22}

The basic lesion is the granuloma, quite variable in number and in various evolutionary phases, indicating that eggs reach the lungs in successive periods. In this report, old granulomas were numerous, most at the stage of fibrosis, in the lumen and adventitia of small arteries.¹⁷

The eggs, once reaching the myocardium, can produce granulomas similar to those found in other sites. They are rarely observed, broadly in patients with massive infections, with fistulae or arteriovenous communications such as described in two cases out of 10,000 autopsies²³ and in 10 out of 4,416 corpses.¹² There are still reports of eggs and granulomas in the right ventricle associated with mural thrombosis¹ and in the right atrium and ventricle with concomitant endomyocardial fibrosis.²² This alleged relationship was established in 15 Egyptians patients out of 10,000 schistosomal patients examined by ultrasound.¹⁶

Renal alterations, in addition to granulomas described in the interstice, are associated, in general, to deposits in the glomeruli of immune complexes, responsible for membranoproliferative glomerulonephritis or, less often, for lobular glomerulonephritis sometimes accompanied by massive proteinuria in patients with the hepatosplenic form of schistosomiasis. In this report, an egg in the afferent arteriole of the glomerulus was still observed.

The pancreatic location, almost constant in the acute toxemic form,¹⁷ was seen in 61 (11.2%) among 544 cases of schistosomiasis¹² and 47.3% of patients with the hepatosplenic form.¹¹ The location of eggs and granulomas is in the interlobular septa, not infrequently determining its fibrous thickening. The parenchyma destruction is topographically restricted to zones of granulomatous inflammation, and the islets of Langerhans are affected only accidentally. This explains why cases of diabetes mellitus resulting from helminthiasis are rare.²¹ This relationship was not found in other accounts.¹⁵

We highlight the existence of a couple of worms in the peripancreatic vein, which allows considering the hypothesis of egg lying on the spot besides the possibility of embolism. The observation in this report of as an association of locations – myocardium, kidney, and pancreas – is also unusual, which has been noted previously only once.¹²

The intestinal location is part of the schistosomiasis infection. Large number of calcified eggs is not uncommon, especially in polyps in the large intestine. Dead worm lesions appear, observed especially in the lungs, and especially in cases treated with praziquantel or oxamniquine.¹⁴

CONCLUSION

This report presents several associations of extreme rarity observed in a young adult male patient in which hepatosplenic, pulmonary, cardiac, intestinal, renal, and pancreatic alterations stand out. We alert to constant attention on schistosomiasis in endemic regions and its various possibilities of differential diagnosis, potential severity, risk of early death, and limitation in the quality of life.

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