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Intestinal Schistosomiasis - a Case Report

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A 61-year old male, an ex-pilot of an agricultural aeroduster, with a history of a *Schistosoma mansoni* infection following an accident in Sudan in 1986 (an exposure to the contents of a polluted water reservoir) was diagnosed three times due to abdominal complaints. The primary diagnosis was Leśniowski-Crohn's disease. Only three years later was an appropriate diagnosis made based on histopathology of sections of the colon.

Introduction

Schistosomiasis (bilharziasis) is a disease caused by parasites belonging to the *Schistosoma* group. An infection with *Schistosoma* was first described in ancient Egypt in approximately 1550 B.C. [2]. The annual worldwide incidence of *Schistosoma* infections is about 150 - 200 million individuals; almost 250,000 die, mostly in consequence of portal hypertension. Several types of *Schistosoma* are human pathogens, such as *S. mansoni*, chiefly occurring in South America, Central Africa and the Near East and mostly attacking the gastrointestinal tract and the liver; *S. haematobium*, which is endemic in Africa (predominantly in Egypt) and in the Near East and most often involves the urinary bladder; *S. japonicum* encountered in the Far East and Philippines, as well as *S. mekongi* and *S. intercalatum* observed in Africa and Indochina [3, 6, 7]. A retrospective analysis carried out by Dr. Laila Farid in Kuwait in the years 1980 - 1987 demonstrated the presence of *Schistosoma mansoni* eggs in the appendix of 2% of patient population, chiefly coming from Egypt [2]. *Schistosoma* matures and reproduces in human blood vessels. The intermediate host for the *Schistosoma* trematode is a snail living in fresh water reservoirs.

The authors describe a patient infected with *Schistosoma mansoni* and hospitalized three times due to abdominal complaints, in whom only a third biopsy of the colon did allow for the diagnosis of schistosomiasis.

A Case Description

A 61-year Polish male used to work as a pilot of an agricultural crop-dusting plane in Sudan. In 1986, his plane broke down and force-landed in a swamp. Most likely he became infected with *Schistosoma mansoni* at that time.

In September 2000, the patient was admitted to Gastroenterology Ward of Public Health Care Hospital in Szczecin, due to abdominal cramps, diarrheal episodes, pruritus and generalized weakness. Lab tests aiming at detecting *Schistosoma mansoni* eggs and other diagnostic tests indicative of a parasitic infection, such as peripheral blood eosinophil level, were negative. The panedoscopy revealed a hiatus hernia, while colonoscopy demonstrated focal blurring of the mucosal vascular pattern in distal sigmoid and rectal segments, as well as a small polyp (2mm in diameter) in the caecum, which was excised but not referred to histopathology. In view of the clinical suspicion of Leśniowski-Crohn's disease, specimens were collected for histopathology, which confirmed the clinical diagnosis (No A8166/00). In February 2002, the patient again reported to hospital presenting with recurrent abdominal complaints. A repeated colonoscopy demonstrated an increased vascular pattern of the sigmoid and rectal mucosa and surface granulation. Specimens for histopathology were collected and the previous diagnosis was confirmed (No A2367/02). For approximately 2.5 years the patient was treated for Leśniowski-Crohn's disease and he improved. The third hospitalization occurred in January 2003, due to intensified gastrointestinal clinical symptom. Another colonoscopy showed a similar pattern as before and specimens were again collected for histopathology. This time, however, the histopathologist was informed about the possibility of a *Schistosoma* infection in the past history of the patient. Microscopically, the colonic mucosa revealed epithelioid cell granulomas where *S. mansoni* eggs could be seen (No A864/03). Preparations from previous biopsies were reviewed and similar histological lesions were found.

Discussion

In inhabitants of endemic regions schistosomiasis is usually asymptomatic due to their acquired immunity [3, 5].

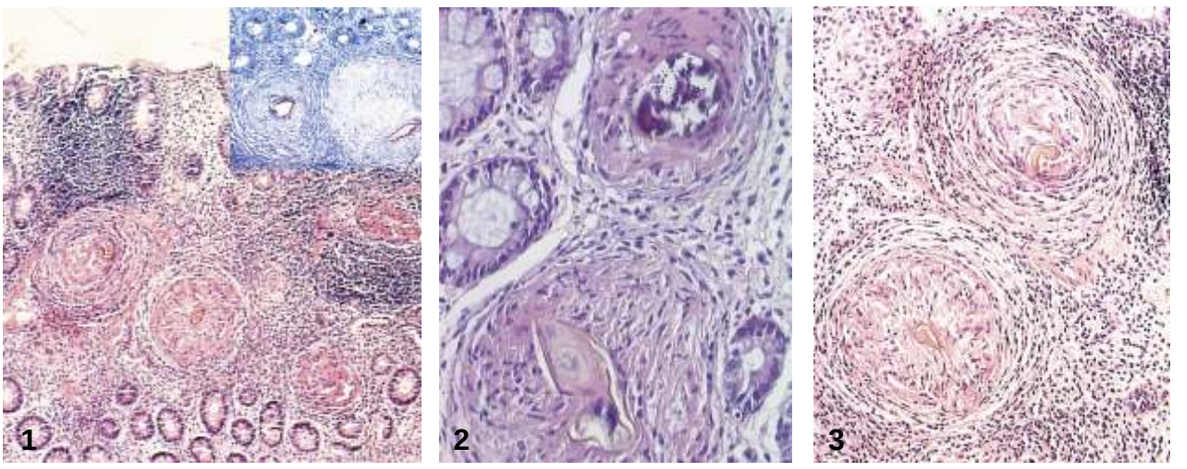

Fig. 1. The lamina propria of the colonic mucosa with an abundant inflammatory infiltration. Note four epithelioid cell granulomas, two of them containing *Schistosoma* eggs. HE. Insert: Ziehl-Neelsen method. The *Schistosoma* egg capsules stain red. Fig. 2. The lamina propria of the colonic mucosa with two granulomas, the central part of which contains *Schistosoma* eggs - one of them is partially calcified. HE. Fig. 3. The lamina propria of the colonic mucosa with two granulomas surrounded by concentric fibrosis. HE.

After the first infection, visitors to these regions develop an acute syndrome (the so-called Kattayama fever), with high temperature, chills, eosinophilia, hepatosplenomegaly, generalized lymphadenopathy and gastrointestinal symptoms. The intensity of intestinal infection varies. The patients complain of abdominal pain, diarrhea and bloody stools. The lesions chiefly involve the rectum and the left part of the colon. The intestinal form of schistosomiasis is associated with an infection with *S. mansoni*, but *S. japonicum* and *S. haematobium* may also trigger similar symptoms. Patients with chronic schistosomiasis manifest mucosal ulceration and luminal strictures that result from the accumulation of granulomas, fibrosis and bilharzial polyps.

In patients with severe forms of intestinal infection the response to conservative therapy is poor and the method of choice is intestinal resection [1].

In the presented patient, the gastrointestinal clinical symptoms persisted for several years. Despite the patient's history indicative of a possibility of a *Schistosoma* infection, laboratory tests failed to detect parasite eggs and thus the clinical presentation suggested the suspicion of Leśniowski-Crohn's disease.

The intermediate host for *Schistosoma* is a fresh-water snail that is infected in its water environment by *miracydia* (transformed eggs). In the snail's respiratory cavity the *miracydia* are transformed into another form, that of *cerkaria*, which in water - due to the proteolytic enzymes that degrade the keratinous layer of the epidermis - penetrate human skin and access the peripheral blood vessels and the lungs to finally colonize the portal or pelvic veins where they mature into adult male and female parasites. The latter produce hundreds of eggs daily. *S. mansoni* lays its eggs in the capillary vessels of the branches of the inferior mesenteric vein; some of these eggs may penetrate the intestinal wall and enter the colon and then they may be excreted with stool [6].

The inflammatory reaction within the colonic wall depends upon the immune status of the patient, the number of *Schistosoma* eggs and the duration of their staying within the host's body. Initially, the patient manifests symptoms of acute intestinal mucosa inflammation: the mucosa is edematous, shows profuse inflammatory infiltration and submucosal hemorrhages. Epithelioid cell granulomas that develop around the eggs constitute the predominant lesion that may calcify; alternately granulomas may encapsulate dead adult parasites that contain *miracydia*. *Schistosoma* eggs are 100 - 180μm in length and approximately 70μm in width. The capsule of an egg is light-brown in color [3]. Apart from epithelioid cells, early granulomas also shows eosinophils, lymphocytes and giant multinucleated cells, while lesions persisting for longer periods show concentric fibrosis.

Epithelioid cell granulomas form in all intestinal wall layers, mostly in the lamina propria of the mucosa and in the serosa. Mature parasites can sometimes be found within the mesenteric veins. They do not evoke any inflammatory reactions.

The histopathological pattern of enteritis triggered by *S. mansoni* should be differentiated from other diseases in the course of which epithelioid cell granulomas develop, mostly from Leśniowski-Crohn's disease and tuberculosis, as well as *Yersinia* infections.

Macroscopically, Leśniowski-Crohn's disease is characterized by skip lesions, the involvement of the entire intestinal wall thickness and the presence of ulceration. Nevertheless, in Leśniowski-Crohn's disease, no such thickening and stiffening of colonic wall is noted as it is the case in the small intestine. In addition, in approximately 20% of cases the rectum shows superficial ulceration and numerous

inflammatory pseudopolyps. Similar lesions are encountered in intestinal schistosomiasis, but without the characteristic skip lesions.

In spite of the fact that the intestinal form of *Mycobacterium tuberculosis* infections is presently rarely encountered, an increasing significance is ascribed to infections with *Mycobacterium avium*, which develop in patients with impaired immunity. Macroscopically and radiologically, tuberculous lesions of the colon resemble the lesions seen in schistosomiasis and Leśniowski-Crohn's disease. Yet usually the former are accompanied by evident mesenteric lymph node enlargement. Lesions resulting from an *Yersinia* infection mostly involve the ultimate segment of the small intestine and the initial segment of the colon, as well as the appendix. Macroscopically they show no major differences as compared to the above-mentioned diseases.

Histologically, the common feature of schistosomiasis, Leśniowski-Crohn's diseases, tuberculosis and *Yersinia* infections is the presence of epithelioid cell granulomas. In the case of schistosomiasis, the granulomas develop in all the layers of the intestinal wall and they can be easily seen in mucosa sections (Fig. 1). The fundamental feature that differentiates granulomas in schistosomiasis is the presence within their center of an egg or a dead parasite; they often undergo retrogressive changes and become calcified, what may hinder their identification (Fig. 2). In addition, granulomas may be surrounded by concentric fibrosis (Fig. 3). In Leśniowski-Crohn's disease such granulomas do not constitute a common phenomenon and they are smaller as a rule. Granulomas are seen in as few as 15 - 36% of colonoscopic biopsies. They usually are not necrotic (sarcoidosis-like granulomas), with the exception of granulomas situated in the serosa - that may undergo necrosis, most likely due to fistula and abscess formation in the adjacent tissues. In Leśniowski-Crohn's diseases, on the other hand, the resected material contains granulomas in 50 - 80% of cases. Later into the course of the disease, the lesions manifest various degrees of hyalinization and fibrosis and involve the entire thickness of the intestinal wall. In intestinal tuberculosis one observes granulomas with distinct caseous necrosis in the central part. In addition, staining by Ziehl-Neelsen method reveals the presence of Koch's bacilli. Caseous necrosis of granulomas is especially intensified in the enlarged mesenteric lymph nodes [7]. In *Yersinia* infections, apart from clearly visible mesenteric lymphadenopathy, and shallow ulcerations, also granulomas may appear, as a rule surrounding microabscesses.

In spite of schistosomiasis being the second after malaria most common infectious disease worldwide, it is relatively rare in Poland. In view of its clinical similarity to various other diseases, the disorder may cause diagnostic errors. This is why a good collaboration between a clinician and a pathologist is of importance, thanks to which clinical information is conveyed that is of great significance in diagnosing such rare tropical diseases as various types of schistosomiasis.

The diagnosis of schistosomiasis lies in detecting *Schistosoma* eggs in the stool, urine or in biopsy material collected from the colon or urinary bladder. Positive serology in itself does not constitute a sufficient foundation for initiating the treatment, but it should prompt a search for live eggs, what - when such eggs are found - justifies the introduction of therapy [1, 4].

References

1. *Berkow R, Flecher A:* Infectious diseases. In: Merck Manual of Diagnosis and Therapy. Urban and Partner 1995, pp 278-279.
2. *Farid L*: Schistosomatosis in Kuwait: A study of some controversial aspects, pathological findings and clinical manifestation. Szczecin 1988 (praca doktorska).
3. *Fenoglio-Preiser CM, Noffsinger AE, Stemmermann GN, Lantz PE, Listrom MB, Rilke FO:* Non-neoplastic lesions of the colon. In: Gastrointestinal Pathology. An Atlas And Text. Lippincott-Raven Publishers 1999, pp 763-908.
4. *Hank C, Verbecke CS, Ltorm S, Junghanss T, Singer MV:* Chronic abdominal pain and eosinophilia in a young African patient. Gastroenterology 2000, 38, 799-802.
5. *Henderson GS, Nix NA, Montesano MA, Gold D, Freeman GL, McCurley TL, Colley DG*: Two distinct pathological syndromes in male CBA/J inbreed mice with chronic Schistosoma mansoni infections. Am J Pathol 1993, 142, 703-714.
6. *Samuelson J*: Infection diseases. In: Robbins Pathologic Basis of Diseases. Cotran RS, Kumar V, Collins T, eds. W.B. Saunders Company 1999, pp 329-402.
7. *Si-Chun Ming, Goldman H*: Ulcerative colitis and Crohn's disease. In: Pathology of the Gastrointestinal Tract. Williams and Wilkins & Waverly Company 1998, pp 669-702.

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