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H. Tesluk

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SEGMENTAL ILEITIS DUE TO SCHISTOSOMIASIS MANSONI

H. TESLUK, M.D.*

This material is being presented for several reasons. Schistosomiasis is an unusual disease in this part of the world and opportunities to study its pathology are rare. Furthermore, the particular complication exhibited by this patient is uncommon for schistosomiasis. Lastly, the clinical and pathological features provide a situation of interest from a standpoint of differential diagnoses. No effort will be made to review the life cycle of the parasite since it is ably presented in numerous sources.1

The patient K.M. is a 28 year old Arabian who came to live in this country fairly recently. He was seen for the first time at Henry Ford Hospital on December 18, 1952 with the chief complaint of abdominal pain. His pain had been present for about four weeks and appeared worse after eating. There was slight nausea but no vomiting. Physical examination showed no unusual findings. Laboratory studies disclosed 4 ++ cephalin and thymol flocculations, but other routine tests were normal. Biliary drainage produced normal appearing bile. However, microscopic examination disclosed ova of the Schistosoma mansoni type. Stool examinations were carried out and the ova seen again, together with Trichuris trichiura. It was decided that the picture was due to schistosomiasis and fuadin therapy administered.

During subsequent visits in the outpatient department a 2-3 cm. mass was noted in the right lower abdomen and a smaller mass on the left side. The liver was slightly enlarged and an x-ray showed a suggestion of fluid levels in the small bowel suggesting early intestinal obstruction.

The abdominal cramps recurred and were especially severe after meals. Again there was no vomiting and no diarrhea. Some weight loss had occurred. Because of the symptoms he was readmitted January 1, 1953. There was a tender mass to the right of the umbilicus. Laboratory studies were normal except for mild anemia. X-ray studies showed multiple polypoid defects of the terminal ileum.

He was again followed as an outpatient, but because of persistence of pain and beginning abdominal distention it was decided to readmit the patient for exploration of the abdominal mass. At operation (July 27, 1953) a huge mass of enlarged mesenteric lymph nodes was encountered in the right lower abdomen, and the terminal ileum was thickened and indurated with almost complete obstruction. The terminal ileum and ascending colon were resected and bowel continuity restored. Postoperatively the patient did well. On August 10, 1953 he was seen again because of persistent epigastric pain. This was thought due to nerve involvement in the operative scar. Repeat x-ray studies showed a well functioning anastomosis.

*Senior Associate in Pathology, Dept. of Laboratories.
Examination of the specimen removed at operation showed the mesenteric nodes to be greatly enlarged and firm. There was some fusion of adjacent nodes. On section they were dense grey white with scattered yellow areas. The terminal 6 cm. of the ileum showed a partial obstruction due to an edematous, polypoid swelling of the mucosa which was sharply localized and ended abruptly at the ileocecal valve. (Fig. 1). There was only a small superficial ulceration. The bowel appeared pale and edematous rather than hemorrhagic. In many ways the gross picture simulated regional enteritis.

Microscopy study showed an intense infiltration of the mucosa with macrophages and scattered about were numerous abscesses in the center of which could be seen ova of the schistosoma worms in varying stages of preservation. (Fig. 2-3.) Eggs which had been present a longer time were surrounded by macrophages and giant cells. (Fig. 4.) The inflammatory process seemed out of proportion to the relatively few ova present and produced a great increase in thickness of the mucosa and submucosa. The cellular reaction consisted predominantly of macrophages together with a few lymphocytes and eosinophiles. The muscularis of the bowel was normal but heavy infiltrations were found between the muscle layers and in the serosa. The appendix and colon showed only a hyperplasia of preexisting lymphatic tissue. Sections from the mesentery demonstrated marked fibrosis and a granulomatous reaction. The circumscribed granulomas were composed of macrophages but giant cells were not present. Frequently there was central necrosis and neutrophiles were fairly abundant. Usually wide bands of fibrosis surrounded these granulomata. They had completely replaced the lymph nodes of the mesentery. No ova could be found in these areas. Another prominent feature was the heavy accumulation of lymphocytes along the course of lymphatic channels. There was also considerable edema present. The smaller veins often showed an obliterative proliferation of their intima. (Fig. 5.) In two of the larger veins there were mature worms. (Fig. 6.) These showed a moderately thick integument and the alimentary tract contained depositions of brown pigment. Periodic acid leucofuchsin stains stained the ova in the mucosa brilliantly and were of considerable help in locating these structures.

**DISCUSSION:**

There are numerous important differences in the diseases caused by the three species, *hematobium*, *mansoni* and *japonicum*. Our patient contracted his infection in Arabia where both the *mansoni* and *hematobium* are found. All three are characterized by the release of ova from adult worms inhabiting venules and in all three the lesions are due to extravasation of ova into tissues. The adult worms are apparently well tolerated. In hematobium infections the adults occupy veins in the pelvis, especially those of the bladder, and most of the lesions involve the urinary tract. In japonicum and mansoni infections, the adults are found in branches of the portal system, and the intestinal tract and liver are the organs which show the most damage. The japonicum infection is more severe due to production of larger numbers of ova. Mansoni infections are usually milder and more protracted. The intestinal lesions are ordinarily encountered in the large
bowel and are characterized by ulcerated polypoid lesions. These are also more severe in japonicum infections. In both diseases the hepatic lesions are more important and most cases die due to hematemesis and cirrhosis. Excellent descriptions of the lesions encountered in mansoni infections are found in Koppisch's classic studies on the disease.²

Our case is unusual because of the curious segmental localization of the intestinal lesions in the terminal ileum and because of their severity which resulted in partial obstruction of the bowel. There is evidence of considerable liver damage on the basis of liver function tests but no tissues were available for study. The studies in Puerto Rico demonstrated that in minimal cases only the liver and rectum were involved and that later, the adults move to higher radicles in the portal system with involvement of mesenteric nodes and small bowel. There were no cases in their series showing such marked localization of the inflammatory polyps in the terminal ileum with resultant partial obstruction. In general lesions of the small bowel were quite minimal in character in their cases except for sub-peritoneal nodules. None were of any clinical significance.

The histopathological features are beautifully described and interpreted in Koppisch's work and no additional information can be added. It is remarkable how few ova were encountered in this lesion considering the bulk and intensity of the inflammatory reaction. The ova elicit an acute inflammatory process at first which is followed by the appearance of macrophages and endothelial cells. Destruction of the embryo results ordinarily and the inflammatory granuloma is encapsulated and fibrosed to form pseudo tubercles. Local sensitization of tissue to toxic factors produced either by the worms or following destruction of the eggs must play an important role in view of the marked inflammatory response. Koppisch points out that the frequent occurrence of necrosis in the center of the pseudo tuberculomas also points to the possibility of an allergic phenomenon.

One feature that is not stressed in previous descriptions is the intensity of the reaction involving lymphatics. Everywhere there is marked accumulation of lymphocytes and plasma cells in and around lymphatics again indicating a response to immunological stimuli. The smaller venules frequently showed proliferation of intima even to the point of occlusion. These are lesions which occur in other granulomatous diseases of the small bowel, especially tuberculosis and terminal ileitis. The differential points between the inflammatory picture in these conditions is clearly discussed by Rappaport.³ It is of considerable interest that many of the features noted in this case, whose pathogenesis and etiology are known, occur in other diseases. The early changes in regional enteritis closely parallel the gross features of this case. Microscopically the same edema and inflammatory lymphangitis is seen but in regional enteritis the proliferative response consists predominately of a hyperplastic lymphadenitis in which granulomatous foci are seen. Similar endophlebitic lesions are also seen and are well illustrated in Rappaport's article. These points of similarity suggest that in regional enteritis the sensitivity or allergic reaction of tissue may play an important part in producing the inflammatory reaction.

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SUMMARY

A case of Schistosoma mansoni infestation is presented which developed the unusual complication of partial intestinal obstruction necessitating resection of the terminal ileum. The pathology of the disease is briefly reviewed and attention drawn to the role of tissue sensitivity in the inflammatory reaction.

Fig. 1—Terminal ileum and caecum. The inflammatory thickening of the mucosa is sharply segmentalized to the terminal ileum.

Fig. 2—Ovum being dislodged into lumen of intestinal gland.
Fig. 3—Ovum with embryo still intact. There is beginning encapsulation.

Fig. 4—Ovum undergoing destruction with foreign body giant cell formation.

Fig. 5—Endophlebitis obliterans together with lymphangitis.

Fig. 6—Adult worms within mesenteric veins.
REFERENCES


