

SCHISTOSOMAL APPENDICITIS IN THE EASTERN PROVINCE OF SAUDI ARABIA: A CLINICOPATHOLOGICAL STUDY

Abdul-Wahed N. Meshikhes, FICS, FRCSI; C.J. Chandrashekar, MD; Qassim Al-Daolah, MBBS; Osama Al-Saif, MBBS; Abdul-Salam Al-Joaib, MBBS; Saed S. Al-Habib, MBBS; Ramadhan A. Goma, FRCSEd

Background: In cases of schistosomiasis, the appendix is commonly infested. It is not known if this is a predisposing factor for appendicitis, or a mere coincidental histological finding.

Patients and Methods: A total of 56 patients (51 males and 5 females) underwent appendectomy for schistosomal appendicitis over a 10-year period at Dammam Central Hospital. The histological slides of 41 of the patients (73.2%) were retrospectively studied.

Results: The highest incidence of schistosomal appendicitis was recorded in the 21-40-year age group. Ova were seen in the submucosal layers of all the excised appendices. The most common tissue responses were submucosal fibrosis (92.7%) and eosinophilia (87.8%), followed by the presence of suppurative inflammation (80.5%). Granulomatous reaction was evident in only 13 cases (31.7%). A striking feature was atrophy of submucosal lymphoid follicles in 70.7% of the cases. Hyperplasia of lymphoid follicles and serosal granulomas were rare (2.4%). Similar tissue responses were histologically seen in four normal appendices examined.

Conclusion: Appendiceal infestation may predispose to appendicitis in the majority of affected cases, but in others, it may well be a mere coincidental histological finding. However, preoperative knowledge bears no clinical significance and does not alter management.

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Key Words: Schistosomiasis, infestation, acute appendicitis, appendectomy.

The increasing incidence of schistosomal infection is giving rise to a major health problem, especially in endemic areas of Africa, the Middle East and South America.^{1,2} The pathological changes of the disease result from the development of a proliferative granulomatous response to the presence of eggs, which are deposited in the submucosa and the mucosa of the rectum and colon, especially the ileocecal region. As eggs are swept back in the portal blood, they become entrapped in the liver, giving rise to a similar granulomatous response.¹

The appendix is the site of ova deposition in 65% of patients infested with schistosomiasis,³ and appendicitis may be the first clinical manifestation of schistosomal infestation. In such patients, it is hard to determine if the presence of these proliferative lesions is a causative factor, or a mere coincidental histological finding. We report here our experience with schistosomal appendicitis in the Eastern Province of Saudi Arabia.

From the Departments of Surgery and Pathology, Dammam Central Hospital, Dammam, Eastern Province, Saudi Arabia.

Address reprint requests and correspondence to Dr. Meshikhes: Consultant Surgeon, P.O. Box 18418, Qatif 31911, Saudi Arabia.

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Patients and Methods

Patients presenting with appendicitis who underwent appendectomy at Dammam Central Hospital over a 10-year-period (1987-1996) and who were subsequently diagnosed as having appendiceal schistosomal infestation, were retrospectively studied. The histological slides were retrospectively reviewed by a pathologist (CJC). All patients except one were diagnosed as having acute appendicitis based on clinical findings.

TABLE 1. *The histopathological features of 41 cases.*

Histological feature	Number (%)
Ova	
Mucosa	26 (63.4)
Muscularis propria	27 (65.85)
Submucosa	41 (100)
Serosa	26 (63.4)
Tissue response	
Submucosal fibrosis	38 (92.7)
Tissue eosinophilia	36 (87.8)
Suppurative reaction	33 (80.5)
Granulomatous inflammation	13 (31.7)
Submucosal lymphoid follicles	
Atrophy	29 (70.7)
Hyperplasia	1 (2.4)

Results

There were 56 patients (51 males and 5 females), with a mean age of 30 years (range 11-55 years). Fifty-four patients presented with symptoms and signs of acute appendicitis, one had an appendiceal mass, and one had incidental appendectomy during an elective open cholecystectomy. Thirty-eight of the patients (69.1%) were non-Saudis, with the majority of these (73.7%) being Egyptian workers. The highest incidence was in the 21-30 years (50.9%) and 31-40 years (38.2%) age groups.

Of the 30 patients with documented white cell counts, 16 (53%) had leucocytosis. Blood eosinophilia was recorded in only six patients. All patients underwent emergency appendectomy. Five patients had a macroscopically normal appendix, i.e., no evidence of acute inflammation as seen by the naked eye, three had gangrenous appendicitis, and another three had perforated appendicitis. There were no documented postoperative complications. None of the patients was investigated for evidence of active schistosomiasis postoperatively.

The histological slides of 41 patients were available for retrospective review (Table 1). The submucosal layer contained ova in all cases. An equal distribution of ova was seen in the mucosa, muscularis propria and serosa (64% of cases). The most common tissue responses were submucosal fibrosis (92.7%) and eosinophilia (87.8%) (Figure 1), followed by the presence of suppurative inflammation (80.5%). Granulomatous reaction was evident in only 13 cases (31.7%) (Figure 2). Atrophy of submucosal lymphoid follicles (Figure 3) was present in 29 cases (70.7%), and hyperplasia was very rare (2.4%). Serosal granulomas were noted in only one case.

In the six normal appendices (including the incidental appendectomy), there was no histological evidence of acute inflammation. The slides were not available for review in two cases. The pathological features of the other four revealed ova in the submucosa only (3), ova in all layers of the wall (1), submucosal fibrosis (4), tissue eosinophilia (3), and granulomatous reaction (1).

Discussion

Schistosomiasis is endemic in the southwestern part of Saudi Arabia.^{4,5} The exact incidence of schistosomal appendicitis in our area over the study period of 10 years was difficult to establish. However, the average incidence at Dammam Central Hospital was 1.5%. This is similar to the incidence reported from the Riyadh area⁶ and the agricultural area of Asir.⁷ The latter is considered to have the highest incidence of schistosomiasis in Saudi Arabia.^{4,5} A higher frequency of 6.2% has been reported from Nigeria.²

Approximately 70% of our cases were immigrant workers from countries where the disease is endemic. More than 73% of those immigrant workers were from Egypt, a

country with a high prevalence of schistosomiasis. As the majority of our patients were immigrant workers, follow-up of these patients poses a problem. In our study, the highest incidence of schistosomal appendicitis was in the 21-30-year age group, followed by the 31-40-year group. This is a higher age group than that reported by Duvie et al., who reported a higher incidence in a younger age group (11 to 15 years).² There was a male predominance (10:1), reflecting the occupational hazard of schistosomiasis, as men who work outdoors in endemic areas are more at risk than females, who mostly work indoors. This is consistent with other previous reports on schistosomal appendicitis.⁶⁻⁸ None of our patients had any symptoms, signs or abnormal liver function tests to raise the suspicion of schistosomiasis preoperatively.

It seems that in some patients, the presence of schistosomal ova was a mere coincidental finding, as was indicated by the histology of the normal appendices, including that of the incidental appendectomy. On the other hand, it can be argued that intense deposition of ova in the appendiceal wall sets up a chronic inflammatory reaction, with subsequent appendicitis. The characteristics of chronic appendicitis, such as fibrosis and chronic inflammatory cells, were strongly evident in the studied slides. Moreover, inflammatory stricture formation and subsequent partial appendiceal obstruction may give rise to symptomatic chronic appendicitis. Therefore, in the majority of affected individuals, the presence of schistosomiasis predisposes to appendicitis and is not just a mere histological coincidence. Nevertheless, schistosomal appendicitis is uncommon in Egypt, a fact which does not support this view.

An interesting histological feature in our study was the lack of submucosal lymphoid hyperplasia, and the presence of lymphoid atrophy in the majority of our cases. This is in contrast to other previous studies which revealed equal incidence (30.7%) of atrophy and hyperplasia of the submucosal lymphoid tissues.⁸ The significance of this is unknown but it warrants further investigation. The earliest histological features of schistosomal appendicitis are granuloma formation (Figure 2) and eosinophilic inflammatory infiltrates (Figure 1). Granulomas are most commonly found in the serosa and submucosa. This is later replaced by submucosal fibrosis with hyalinization.⁹ Submucosal granulomas were present in one-third of our cases, while serosal granulomas were encountered in only one case. Submucosal fibrosis was the most common response encountered (92.7%) in the studied specimens, reflecting the chronicity of the disease in our patients.

Unfortunately, none of our patients was investigated for active schistosomiasis by examination of fresh rectal biopsy for viable ova under direct microscopy. Long-term follow-up was virtually impossible, as the majority of our patients were immigrant workers. Therefore, only a small number of the patients received antischistosomiasis therapy, at the discretion of the treating surgeon, as the

majority of them were discharged before histological confirmation of the condition.

We believe that the presence of schistosomal infestation increases the risk of appendicitis in the affected individual, but does not always precipitate it. Preoperative knowledge of the infestation does not bear any clinical significance, as it does not influence the ultimate treatment, which is simply an appendectomy. Nevertheless, lack of knowledge before histopathological confirmation may jeopardize the search for and treatment of the active disease in the absence of follow-up.

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