Katayama syndrome

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ABSTRACT

Two schoolboys from a non-endemic region visited an endemic area of Schistosomiasis in Yemen. They developed cutaneous itching after swimming in infected ponds. Five weeks later both brothers were admitted to the hospital with high fever, abdominal pain, vomiting and diarrhea. The blood results exhibited marked eosinophilia and schistosomal serological test showed an evidence of exposure. Initially, stool examinations were negative, but the sigmoidoscopy was suggestive for acute bilharzial colitis and the biopsy showed schistosoma ova; a finding consistent with Katayama syndrome. Both patients treated with Praziquantel, and they became asymptomatic, and the eosinophilia normalized. This report shows the importance of endoscopic and histological examination in suspected acute colonic schistosomiasis, which allows early treatment and avoids possible complications.

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Schistosomiasis, also known as bilharziasis or snail fever, is an infectious disease, which was described 1900 BC and still is a major public health problem in tropical and subtropical regions. More than 200 million people are infected worldwide.[1,2] The eradication of schistosomiasis has been difficult, despite the recent development in research, thus, early diagnosis and treatment will prevent serious complications.[3] There are 5 species of schistosomiasis infecting humans, mainly *Schistosoma (S) Mansoni, S. Japanicum, S. Hematobium, S. Intercalatum and S. Mekongi.* Acute infection is often asymptomatic, but can present with non-specific flu like symptoms or in extreme cases as the potentially fatal form such as Katayama syndrome. This is a severe febrile illness accompanied by abdominal pain, diarrhea, cough, hepatosplenomegaly and eosinophilia, which may occur 4-6 weeks following exposure. If the disease is not early recognized and treated, it will gradually lead to chronic schistosomiasis with its consequences.[4,5]

Case Report. Two boys from the Eastern region of the Kingdom of Saudi Arabia, aged 11 and 13-year-old, were admitted to King Faisal University Hospital, Al-Khobar. Both patients had fever, abdominal pain, vomiting, diarrhea and hepatosplenomegaly. Further information revealed history of journey to an endemic area of schistosomiasis in Yemen and swimming in infected ponds. Both brothers gave history of swimmers itch and 4 weeks later they developed the other symptoms, which brought them to emergency room. During hospitalization, they developed episodes of loose motions with mucous and occasional blood. Patients remained ill with fever and severe abdominal pain, which required surgical consultation. Initial laboratory investigations in both patients showed leukocytosis of 29 and 40,000 with marked eosinophilia of 39 and 80% and erythrocyte sedimentation rate was 40 and 60 mm in the first hour. Liver functions in both patients remained

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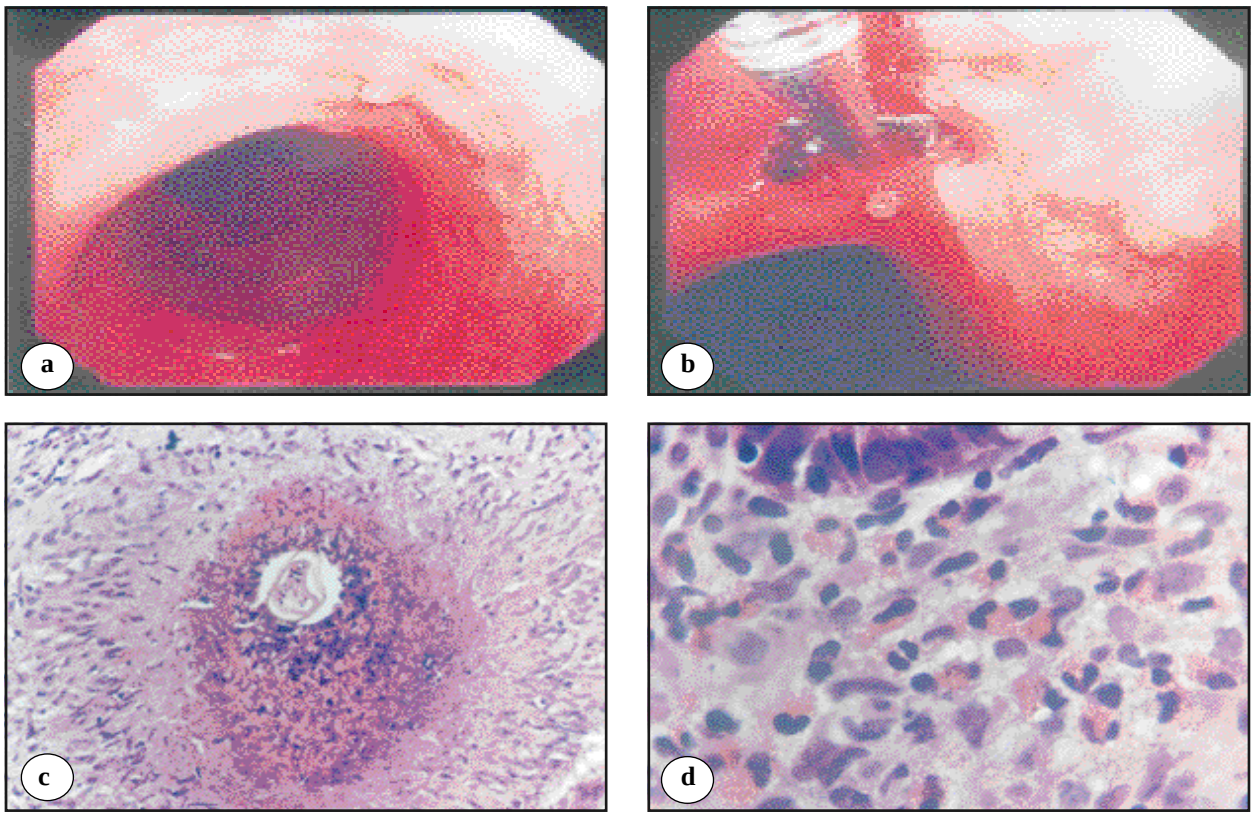

Figure 1 - Sigmoidoscopy showing **a and b)** small polyp (nodule) which was biopsied. **(c)** Epithelioid cell granoloma with central fibrinoid necrosis around a viable schistosoma ovum. Hematoxylin and Eosin staining x 250. **(d)** Cellular inflammatory reaction around schistosoma ova. High magnification showing numerous eosinophils. Hematoxylin and Eosin staining x 1000.

normal, and the abdominal sonography confirmed the hepatosplenomegaly. Repeated examination of the feces revealed the presence of *Schistosoma mansoni* ova in one of the patients. The sigmoidoscopy of both patients showed petechial lesions and small nodules with evidence of an acute colitis **(Figures 1a & 1b).** Biopsy from the lesions revealed signs of acute bilharzial colitis with schistosoma eggs **(Figures 1c & 1d)**. The serology indirect hemaglutination (IHA) was found to be significantly raised to 8192 in both patients. Other investigations, such as electrocardiogram, chest x-rays, blood and stool cultures malaria, serological tests for Amoeba, Salmonella, brucella, hepatitis, anti streptolysin-o-titer, monospot test, as well as gastroscopy were normal. Bone marrow aspiration showed adequate iron storage and sideroblasts with marked increase in eosinophilia and their precursors. There was no bone marrow infiltration or infection. Both patients were given a single dose of praziquantel 40 mg per kg body weight. 3-5 days later patients showed significant improvement. By the end of the first week all symptoms improved and patients were discharged. Two months later, the patients were asymptomatic, liver, spleen and eosinophilia were back to normal. After 4 months, the second dose of praziquantel was prescribed.

Discussion. Katayama syndrome was first described in 1847, in a village in Hiroshima, Japan. It usually occurs 4-6 weeks after infestation; it is an allergic manifestation, developing, in non-immune persons who are exposed for the first time to schistosomal infection. Manifestations include fever and eosinophilia. It may be confused with other acute febrile illnesses such as gastroenteritis, Enteric fever malaria and brucellosis. The diagnosis of this syndrome is based on a history of recent exposure and the presence of *S. ova* in the feces. The demonstration of *S. ova* in the stool is not always easy since the ova passage into the intestinal lumen is usually inconstant, reduced and intermittent. Repeated examination and concentration technique of stool may improve the sensitivity. Biopsy of the rectum for ova is more sensitive than stool examination.[6-8] Histological examination typically

shows well formed granulomas with mixed inflammatory response, including eosinophils. The high yield rate of colonic biopsies depends on obtaining multiple endoscopic biopsies from several sites. Positive schistosomal serological tests early in the disease may help in the diagnosis before the appearance of the ova in the feces. In Katayama syndrome, schistosoma antibody serology titre is usually very high as was seen in our patients.[9] In principle, schistosoma serological tests indicate the presence of antibody whether recent, inactive or well-established infection and cannot differentiate between those conditions. Its main rule remains as an epidemiological value.[10,11] Clinical and immunological manifestations of Katayama fever are similar to those of serum sickness, suggesting that immune complex is important in the pathogenesis of this acute disease.[12,13] However, clinical manifestation may vary widely.[3,14,15] Our patients were extremely symptomatic with impressively elevated leukocytosis and eosinophilia, however, the liver functions remained normal. The role of anti schistosomal drugs, in altering the acute course of the disease is controversial.[3,12] However, both our patients by the end of first week of therapy were feeling well and ready for discharge. While in untreated cases, symptoms can last for months.

In conclusion, we found, that it is essential to be aware that sigmoidoscopy with biopsy is superior to routine stool examination and even more important to recognize and treat. The early treatment of Katayama syndrome, alters the course of the acute illness and will halt the progress of the disease; also failure to do so will eventually lead to the individual patient sustaining damage, chronicity and serious complication.

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