CASE REPORT

Sweet Syndrome in a Case of Schistosomiasis—A Rare Complication of Rare Disease: A Case Report

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Abstract

Introduction: Schistosomiasis, also known as bilharziasis is a parasitic disease caused by blood flukes (Trematode worms) of the genus *Schistosoma*. Schistosomiasis is prevalent in tropical and subtropical areas, especially in poor communities without access to safe drinking water and a lack of sanitary facilities. It is rare in the Indian subcontinent. Sweet's syndrome, also known as acute febrile neutrophilic dermatosis, is a rare inflammatory condition. It is considered to be the prototype disease of neutrophilic dermatoses.

Case description: Here we report a case of urinary schistosomiasis along with sweets syndrome in a case of 56 years old male residing at Digha, West Bengal. Although it is very rare in our country, it is probably the second case report of schistosomiasis in India after Maharashtra. Again, schistosomiasis complicated by sweet syndrome was never been reported worldwide.

Conclusion: This case report should aware the physician treat any patient with fever with hematuria, particularly living in the coastal area and/or history of travel to the coastal area in our country or abroad.

Keywords: Bilharziasis, Case report, Schistosomiasis, Sweet syndrome.

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Case Description

A 56 years old male, non-diabetic, non-hypertensive was admitted under our care with a history of intermittent fever for 4 days followed by erythematous plaques with psuedovesiculation over the trunk, lower limb, and upper limb, (Fig. 1) arthralgia, redness of the eye, shortness of breath, malaise, abdominal pain, and terminal hematuria. Fever was evaluated and was negative for dengue NS1, malaria, leptospira, scrub typhus, and typhoid. Total bilirubin came – 4.3 mg/dL, LDH – 733 U/L, Hb was – 6.9 gm/dL indicating microangiopathic hemolytic anemia. D dimer and FDP were raised – 6.04 mg/L FEU and 8433.09 µg/mL respectively, along with hematuria pointing to suspected disseminated intravascular coagulopathy, and injection unfractionated heparin 5000 unit subcutaneously was given for 5 days. Ferritin being an acute inflammatory marker was also raised to – 1085 ng/mL. Contrast-enhanced computed tomography (CECT) scan of the thorax and abdomen was normal. Serum urea and creatinine were raised suggestive of acute kidney injury and the patient subsequently underwent hemodialysis 4 times with a serial decrease in serum urea and creatinine. Skin biopsy from lesions for histopathological examination revealed mild hyperkeratosis of the epidermis. Dermis shows a collection of neutrophils in the papillary dermis and perivascular neutrophilic infiltration in the reticular dermis - suggestive of Sweet's syndrome (Fig. 2). As part of the evaluation for Sweet's syndrome, PET – a CT scan of the whole body was planned but it showed no significant metabolically active abnormal malignant focus. The ultrasonography of the whole abdomen did not reveal any abnormality. Antinuclear antibodies (ANA) profile study was sent but did not show any abnormality.

Urine routine examination of revealed RBC: 10–15/high power field, epithelial cells: 3–5/high power field, pus cells: 3–4/high power field; wet mount field revealed the presence of non-operculated ova with terminal spine, morphologically resembling *Schistosoma haematobium* (Fig. 3). Urine culture and blood culture showed no

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Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

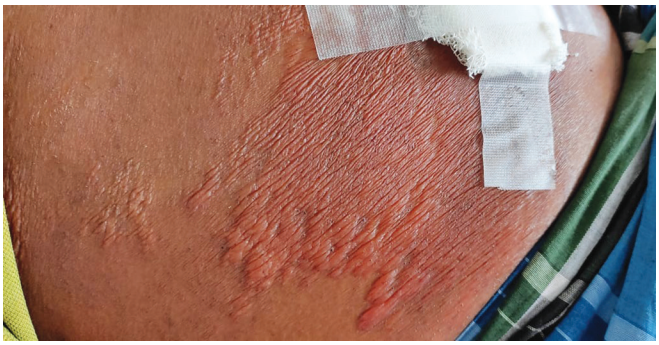

Fig. 1: Erythematous plaques with psuedovesiculation

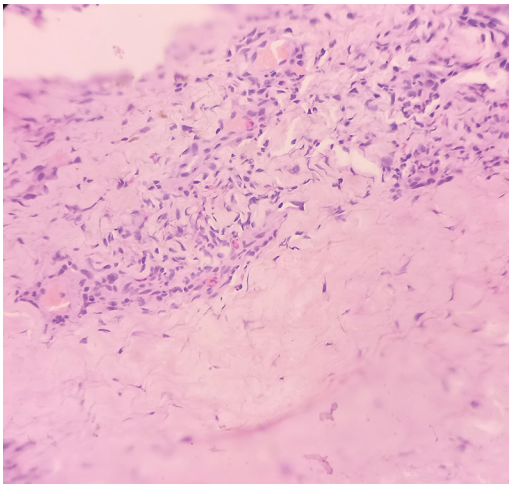

Fig. 2: Collection of neutrophils in papillary dermis and leukocytoclastic vasculitis

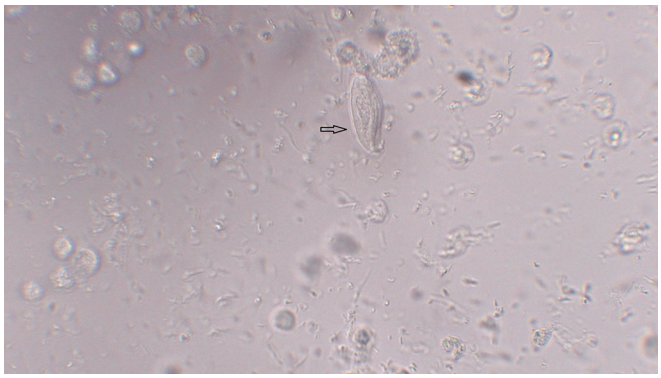

Fig. 3: Arrow shows nonoperculated ova with terminal spine, morphologically resembling *Schistosoma haematobium*

growth. A 24-hour urine sample, after centrifugation, ZN stain was done and it came negative for Tuberculosis. A repeat urine examination was done that confirmed the ova of *S. haematobium*. Polymerase chain reaction (PCR) based study was not done as it was unavailable. Ova of *S. haematobium* was not found after a routine examination of stool. The patient was treated with Tablet Praziquantel 1,200 in two divided doses on a single day. The patients showed gradual improvement and disappearance of the skin lesions and repeat urine microscopy after 3 days did not show any ova or parasite.

Discussion

Schistosomiasis or Bilharziasis is a fresh waterborne parasitic disease caused by the Schistosoma genus of Trematoda class commonly known as blood flukes.[1] Urogenital schistosomiasis is caused by *S. haematobium* and intestinal schistosomiasis is caused by *S. guineensis, S. mekongi, S. japonicum, S. mansoni, S. intercalatum*.

The female fluke releases 300–3500 eggs daily into the blood. These numbers vary according to species. The eggs are evacuated through feces and urine. The eggs hatch in the freshwater. It releases ciliated larvae that enter into the intermediate or snail host. The cercaria or the fork-tailed larvae, after emerging from the freshwater snail, penetrate the skin and soft tissue of mammals and enter into the blood circulation. In this way, they drop their tails.[2] Schistosome worms do not multiply in the host. The infection status is the result of consecutive re-infections, and the most intense infection usually has a higher chance of developing morbidity.[3]

The characteristic sign of urogenital schistosomiasis is painless terminal hematuria. In the advanced stage of disease progression, they damage the kidneys and finally lead to fibrosis of the bladder and ureter and causes bladder cancer.

In human beings, disease progression of Schistosome infections can be divided into three distinct stages: Acute, active, and chronic. Symptomatic acute schistosomiasis, also known as Katayama fever or Katayama syndrome, usually occurs between 2 weeks and 3 months, after exposure to the parasite due to systemic hypersensitivity reaction and immune complex formation which occurs during its passage through the human tissue.[4] In the active stage, it secretes live eggs in stools and urine. Antigenic glycoproteins secreted by schistosome eggs help in their migration from the blood circulation to the lumen of the bladder and intestine by an inflammatory response. This is accompanied by sloughing off of the epithelial surface of the bladder wall which leads to hematuria, thickening of the bladder wall and pseudo polyps in bladder are developed.[5]

"Acute febrile neutrophilic dermatosis" is the original description of Sweet's syndrome by Sweet.[6] It is an inflammatory, non-infectious skin reaction. Clinically it presents with tender, erythematous papules/nodules/plaques/pustules. There are three distinct variants: Malignancy-associated, drug-induced, and classical. Von den Driesch criteria is used for the diagnosis of sweet syndrome.[7]

Detection of Schistosome egg in urine or stool is indicative of acute infection and is the standard diagnostic method. Histopathological examination of urinary bladder specimens or rectal biopsy specimens for eggs of S. haematobium may be used as an alternative diagnostic method.[8]

Schistosoma haematobium has a very low incidence rate in South Asian countries. An endemic focus of *schistosomiasis* of uropathogen was confirmed. In Gimavi Village of Ratnagiri district, Maharashtra, India. This was postulated as a new *Schistosoma* species due to its distinct oval-shaped egg and the absence of the Bulinus species of snail in India as the intermediate host. So *Schistosoma gimvicum* as a new species has been proposed.[9] There has been no case report in the literature on schistosomiasis and Sweet's syndrome occurring together in a patient.

Conclusion

This case report confirmed urinary schistosomiasis caused by *S. haematobium* with typical egg morphology in urine sample alongside Sweet's syndrome diagnosed clinically and by biopsy and histopathological examination. African and Middle east countries are endemic for urinary *Schistosomiasis*. But this case reports shows that the disease can be diagnosed in countries like India with huge population and their travel in foreign countries. This human infection indicates possibility of presence of indigenous snail as an alternate intermediate host of human *Schistosomiasis*. So, diagnosis of such a rare disease in India needs strong mind of clinical suspicion of the disease in physician. Further investigations are suggested on the possible causal relationship between Sweet's syndrome in *Schistosomiasis*.

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