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# A Solitary Nodular Vulvar Lesion

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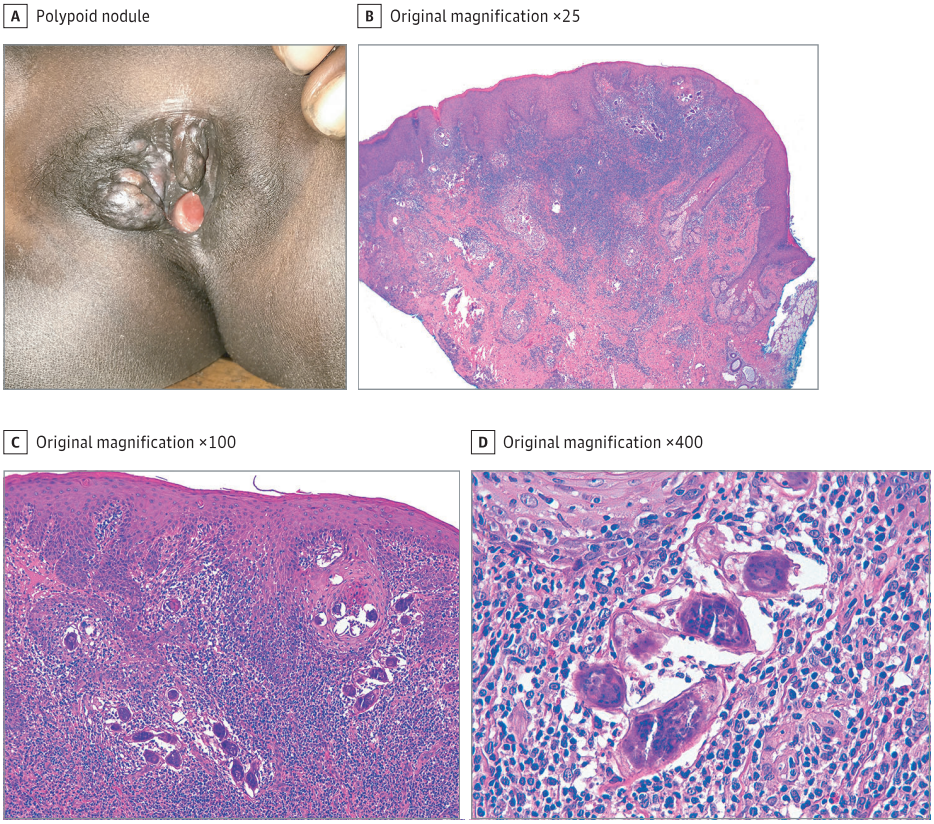


**Figure.** A, Polypoid nodule on the right labium majus obliterating the labium minus. B-D, Hematoxylin-eosin–stained lesional specimens. B, Histopathologic analysis of the nodule showed pseudoepitheliomatous hyperplasia with transepidermal elimination of basophilic oval-shaped structures in the epidermis and a dense acute and chronic dermal infiltrate in the dermis. C, Numerous dermal structures surrounded by a dense acute and chronic inflammatory cell infiltrate with granuloma formation. D, High-power magnification of the oval parasitic structures with a typical terminal spine.

**A girl in her teens** presented with a unilateral itchy nodule on her right labium majus of 4 months' duration. The patient was born and lived in a rural village in Malawi. Her medical and family history were unremarkable. She did not report fever or malaise. Physical examination revealed a polypoid, rubbery tumor measuring 1.5 cm located on her right labium majus, which obliterated the right labium minus (**Figure**, A). She did not present with enlarged lymph nodes, and results of the physical examination were otherwise unremarkable. An incisional lesional biopsy was taken (Figure, B and C).

**WHAT IS YOUR DIAGNOSIS?**

**A.** Onchocerciasis

**B.** Late cutaneous bilharziasis

**C.** Periorificial cutaneous tuberculosis

**D.** Granuloma inguinale

## Diagnosis

**B.** Late cutaneous bilharziasis

## Discussion

Histopathologic examination showed pseudoepitheliomatous epidermal hyperplasia and transepidermal elimination of basophilic oval-shaped structures. A dense acute and chronic inflammatory cell infiltrate composed of neutrophils, eosinophils, lymphocytes, and macrophages with granuloma formation was identified in the dermis. Numerous parasitic structures were observed in the granulomas (Figure, B and C). At high-power magnification, basophilic oval-shaped parasites with a terminal spine characteristic of *Schistosoma haematobium* were identified in the dermis (Figure, D).

Given these histopathologic findings, the diagnosis of late cutaneous bilharziasis (LCB) was made. However, the patient was lost to follow-up. Therefore, no additional tests could be performed to exclude complications (urine or fecal analyses, abdominal imaging), nor treatment given.

Late cutaneous bilharziasis, or late cutaneous schistosomiasis, is a chronic cutaneous infection caused by trematodes of the *Schistosoma* genus. It is the second most common parasitic infection in the world and is more frequent in freshwater-rich areas of the African continent. However, with migratory flows, cases can be observed worldwide.[1] *Schistosoma hematobium* is the most common causative agent. However, infections by *Schistosoma mansoni* and *Schistosoma japonicum* have also been reported.[2] The genitalia are the most common location of LCB, even if extragenital location can also occur. Vulvar involvement, considered to be rare in genital LCB (about 7%-17%),[2] is more frequent in prepuberal age, although it may be underreported.[3]

There are different clinical forms of cutaneous schistosomiasis according to the immune status of the patient and time of presentation after exposure: swimmer's itch, Katayama fever, and LCB. Late cutaneous bilharziasis is produced by deposition of the parasite eggs in the dermis by retrograde migration through portal, perivesical, and mesenteric venules from previous systemic infection. Granuloma formation and transepidermal elimination subsequently occur to eliminate the parasites.[4] Eventually, granulomatous reactions produce clinical manifestations, such as itchy papules, nodular lesions and polypoid tumors. Urinary (in *S hematobium* infection) and fecal (in *S mansoni* and *S japonicum* infection) parasitic discharge and the corresponding signs and symptoms can occur concomitantly.[2] Possible complications of *S hematobium* infection are the development of vesical squamous cell carcinoma, hydronephrosis, infertility, increased transmission of sexually transmitted diseases, and pseudoelephantiasis, among others.[1-3,5,6]

Confirmation of LCB diagnosis requires microscopic detection of eggs in urine, feces or skin; *S hematobium* has a terminal spine, while *S mansoni* has a lateral spine. Enzyme-linked immunosorbent assays for IgG, IgM, and IgE are available and distinguish acute from chronic infection. Onchocerciasis, periorificial cutaneous tuberculosis, cutaneous amoebiasis, and granuloma inguinale are the main differential diagnoses of LCB.

Onchocerciasis is a nematode infection by *Onchocerca volvulus*. It can present with atrophic hypopigmented macules, blindness, and skin nodules called onchocercomas. However, no vulvar onchocercomas have been reported to our knowledge. Biopsy specimens show microfilaria in the dermis. Vulvar periorificial cutaneous tuberculosis is very rare, and is caused by hematogenous or urinary inoculation of *Mycobacterium tuberculosis*. Cases clinically similar to the present one have been described,[7] but histopathological findings were different, showing granulomas with caseation. Cutaneous amoebiasis is an infection by *Entamoeba histolytica*. Vulvar cases are extremely rare and present with verrucous and/or ulcerated plaques through contiguous infection from amoebiasis colitis. *Entamoeba* trophozoites can be observed in biopsy specimens, and they are CD59 positive (N-acetylgalactosamine positive). Granuloma inguinale or donovanosis is a rare infection by *Klebsiella granulomatis*. A hypertrophic form exists, presenting with clinical features very similar to those of the present case. However, Donovan bodies may be found in pathologic studies.

The treatment of choice in LCB is praziquantel, 40 mg/kg, 2 or 3 doses.[8] When fibrotic tissue remains after praziquantel treatment, surgery is indicated.

In conclusion, we present a rare case of vulvar bilharziasis. Early diagnosis requires adequate clinicopathological correlation to prevent complications.


### ARTICLE INFORMATION

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