

From the Dermatologikum Hamburg: Quiz

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The patient

A 7-year-old girl was presented by her parents with a nodule on the right leg. The lesion had been present for a few weeks and had slowly increased in size. At the time of presentation the lesion measured 6 mm in diameter and it was neither painful nor itchy (Figure 1A). Skin examination revealed two additional smaller red papules on the same leg (Figure 1B). A biopsy was taken from the lesion pictured in Figure 1A.

Histopathologic changes are presented in Figures 2A-E. What is your diagnosis? Would you ask for any special stains or other investigations for this patient?

Answer and explanation

The changes shown in Figures 2A-E are those of a sarcoidal granulomatous dermatitis. The disease that first comes to mind when looking at this pattern is sarcoidosis. However,

the clinical presentation with few localized small nodules or papules in a child would be very unusual. Interestingly, nodular aggregations of macrophages in this case are joined by some lymphocytes and few plasma cells, which should always raise the suspicion of an infectious process. Special stains performed in this case included PAS, Ziehl-Neelsen, Fite, and Giemsa, but all of them were negative.

Polymerase chain reaction (PCR) performed on the paraffin embedded specimen, however, revealed leishmania-specific DNA. Sequencing identified *Leishmania infantum* as the causative organism. The lesion and the other two papules were excised and *Leishmania infantum* was also found in these additional specimens.

Leishmania species are transmitted by the bite of certain species of the sand fly [1-3]. Clinically leishmaniasis shows a wide spectrum of clinical manifestations including localized cutaneous lesions, mucocutaneous involvement, or disseminated and potentially lethal infections. The parasites



Figure 1A and B. Red nodule (A) and two red papules on the right leg (B). [Copyright: ©2013 Böer-Auer.]

are mainly found in South Asia, the Horn of Africa, Central and South America and in the Mediterranean region. In non-endemic countries leishmaniasis is a disease of travel-

ers and migrants. While some species of leishmania tend to involve only the skin (e.g., *L. tropica*), others bare the risk of mucocutaneous or systemic infection (e.g., *L. infantum*). Outdoor activities in endemic regions put unaware individuals, especially children, at risk of being bitten and infected. For infection with *L. infantum*, it has been shown that children below the age of 3 years have a higher risk of developing visceral disease compared with older children and adults, a finding that has been attributed to age related differences in T-cell immunity [2]. Therefore, it is important to consider a diagnosis of leishmaniasis in any child that presents with a single or few nodules, especially when lesions are located on uncovered skin areas and when there is a history of travels or residence in endemic areas [3].

Classic histopathological findings of cutaneous leishmaniasis are those of a tuberculoid granulomatous dermatitis with plasma cells and evidence of amastigotes in histiocytes or free in the tissue [4]. Lesions are often ulcerated and crusted. However, it has been demonstrated that cutaneous leishmaniasis may also present with other types of granulomas, including sarcoidal, palisaded, interstitial, and foreign body types [3,5,6]. In such cases, the load of organisms is often low and a specific diagnosis can only be reached with additional culture or with PCR. More

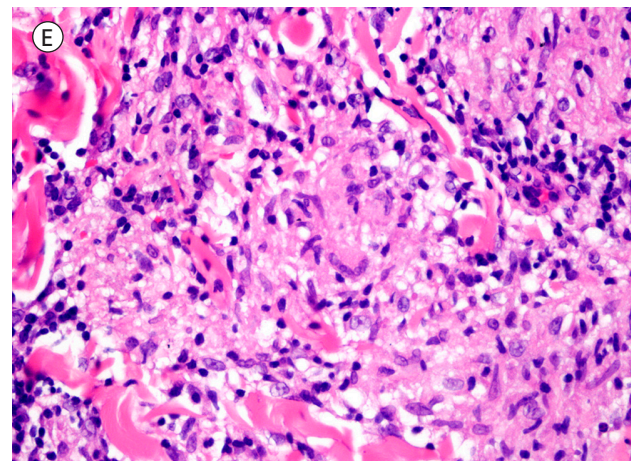
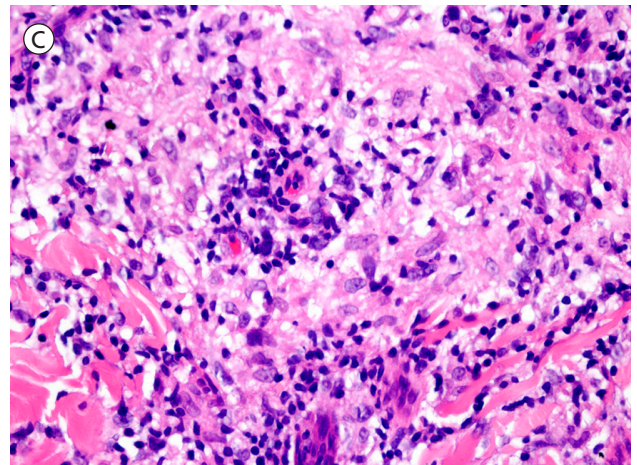
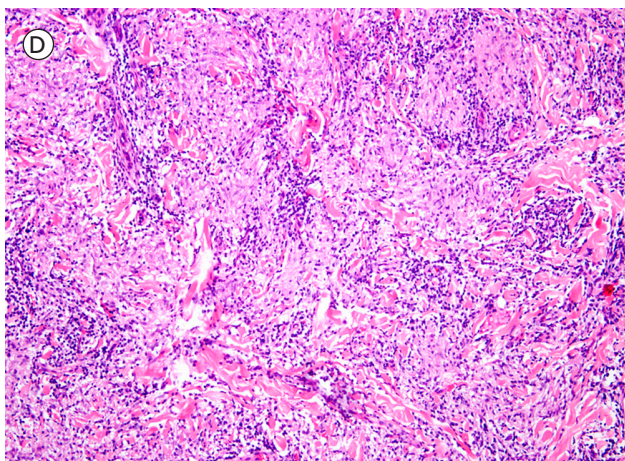
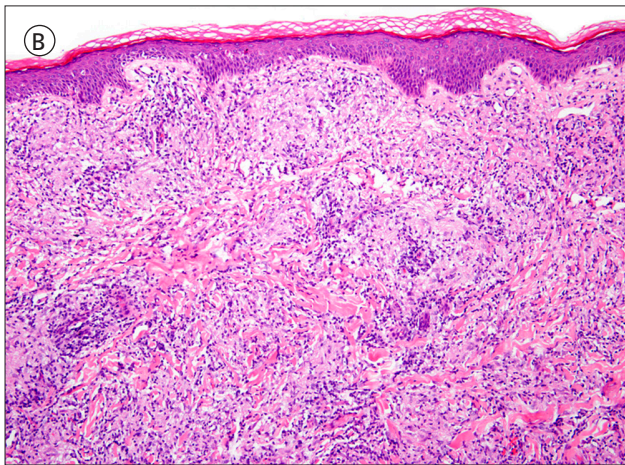
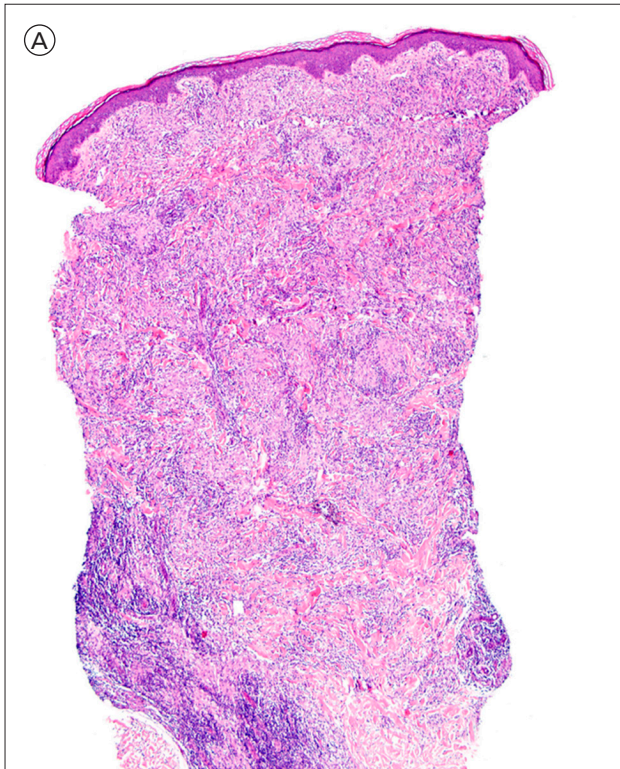


Figure 2A-E. What is your diagnosis? [Copyright: ©2013 Böer-Auer.]

recently, PCR for leishmania specific DNA has become very reliable even when it is performed on paraffin embedded tissue. By way of sequencing, specific subtypes can be identified in such specimens so that culture may not be necessary. Exact typing should be performed because it helps to identify patients at risk for mucocutaneous and systemic infections, and it is also important in order to select the most appropriate treatment [7].

In the patient presented here, no lesions of leishmaniasis could be identified at any other site of the body. Treatment consisted of surgical removal of the lesions and close follow-up, which up to now has been uneventful. The girl had acquired the infection most likely during a vacation to the Mediterranean region.

In sum, in a patient with a sarcoidal granulomatous dermatitis with plasma cells, leishmaniasis should be considered, especially when the clinical presentation is that of one or few lesions on skin surfaces that are usually uncovered in warm climate. If no amastigotes can be identified in routine stains, the specimen should be submitted to polymerase chain reaction for leishmania specific DNA.

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