The best diagnosis is:

- a. cutaneous Rosai-Dorfman disease
- b. granuloma annulare
- c. juvenile xanthogranuloma
- d. necrobiosis lipoidica diabeticorum
- e. necrobiotic xanthogranuloma
Necrobiosis lipoidica diabeticorum (NLD) is a rare granulomatous skin manifestation that is strongly associated with diabetes mellitus. Necrobiosis lipoidica diabeticorum is more common among females and occurs primarily in the pretibial area. \(^1\) Necrobiosis lipoidica diabeticorum may clinically manifest as single or multiple lesions that begin as small red papules and progress into patches or plaques. Lesions ultimately develop into areas of yellowish brown atrophic tissue with central depression and telangiectasia. The etiology of NLD is not completely understood, but it is thought to be a presentation of diabetic microangiopathy.\(^1\) Histologically, NLD demonstrates broad horizontal zones of necrobiosis with a surrounding inflammatory infiltrate that is principally composed of histiocytes but also may contain multinucleated giant cells, lymphocytes, and plasma cells (Figures 1 and 2). Occasionally, sarcoidal granulomas are seen in NLD. There also may be thickening of vessel walls and edema of the endothelial cells.\(^1\)

Cutaneous Rosai-Dorfman disease (RDD) is characterized by the presence of diffuse, large, pale histiocytes (commonly known as Rosai-Dorfman cells) with an admixed infiltrate of lymphocytes and plasma cells (Figure 3).\(^2\) Additionally, Rosai-Dorfman cells display emperipolesis. They stain positively for S-100 protein and CD68 and negatively for CD1a.\(^2\) Clinically, cutaneous RDD has a myriad of manifestations but most commonly presents as cutaneous nodules that can be tender or pruritic. It also may be associated with systemic symptoms. Patients with cutaneous RDD often have an elevated erythrocyte sedimentation rate and concomitant anemia.\(^2\)

Granuloma annulare demonstrates necrobiosis and palisaded granulomatous dermatitis similar to NLD; however, the necrobiotic foci in granuloma annulare usually are more focal than in NLD and typically are surrounded by well-formed palisaded granulomas. There also is an increase in dermal mucin (Figure 4), which can be highlighted on colloidal iron or Alcian blue staining.\(^1\) Granuloma annulare also typically has scattered eosinophils rather than plasma cells as seen in NLD. Granuloma annulare also may present in an interstitial pattern, with scattered histiocytes,
mucin, and eosinophils between collagen bundles. Granuloma annulare clinically presents as variably colored papules arranged in an annular pattern on the distal extremities but also can present as widespread papules or plaques.

Juvenile xanthogranuloma (JXG) is a benign condition typically seen in children that is characterized by the presence of 1 or more pink or yellow nodules, most commonly presenting on the head and neck. Histologically, JXG demonstrates a dermal collection of histiocytes, lymphocytes, eosinophils, and characteristic Touton giant cells, which contain nuclei that are arranged in a wreathlike pattern and exhibit peripheral xanthomatization (Figure 5). The histiocytes in JXG typically stain positive for CD68 and negative for S-100 protein, though occasional S-100–positive cases are reported.

Necrobiotic xanthogranuloma presents as yellowish to brown plaques and nodules most commonly in the periorbital area. Necrobiotic xanthogranuloma is strongly associated with monoclonal gammopathy, typically IgGκ monoclonal gammopathy. Necrobiotic xanthogranuloma is histologically similar to NLD but is distinguished by a nodular pattern of inflammation and the frequent presence of cholesterol clefts (Figure 6).

REFERENCES