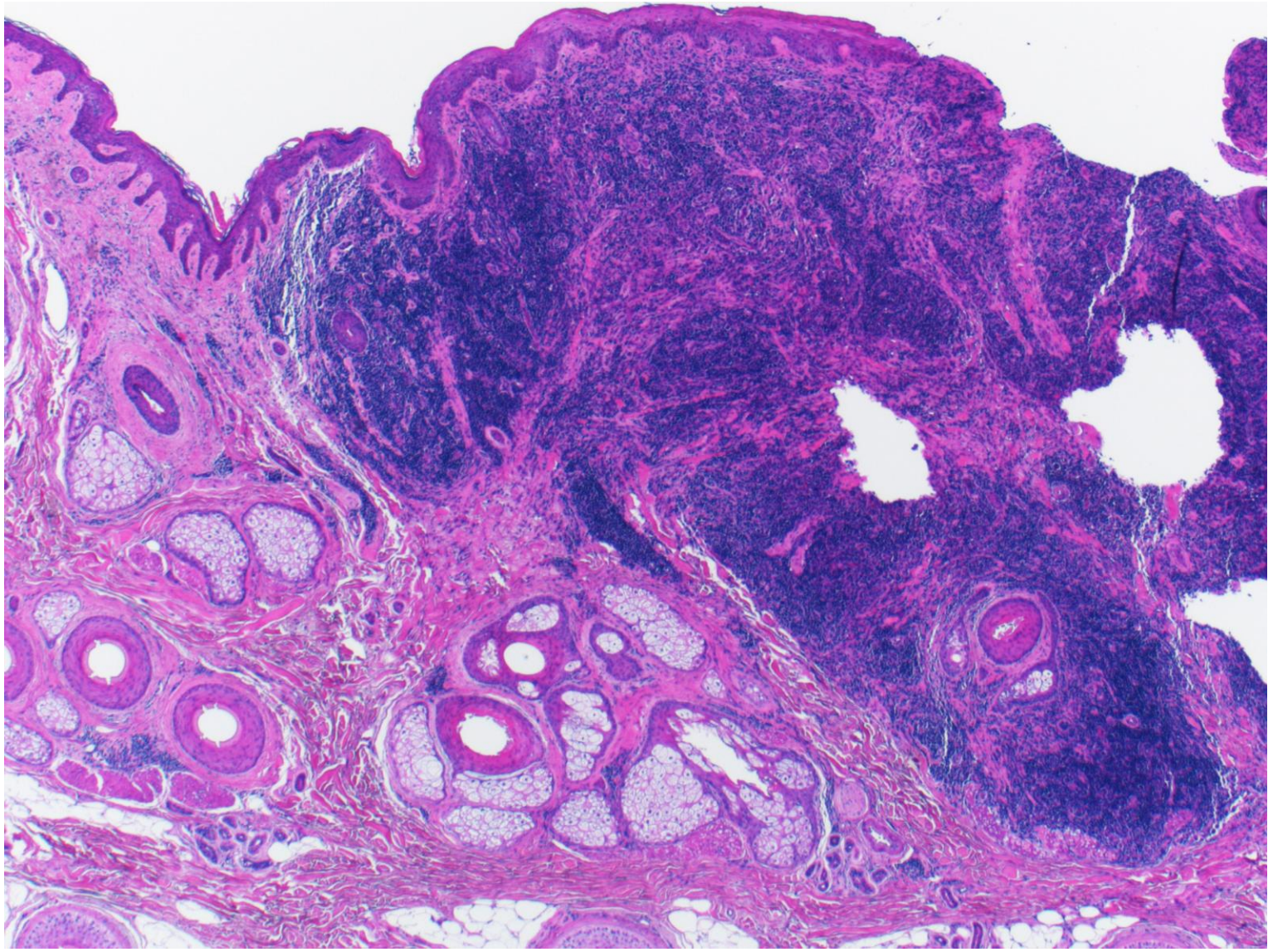
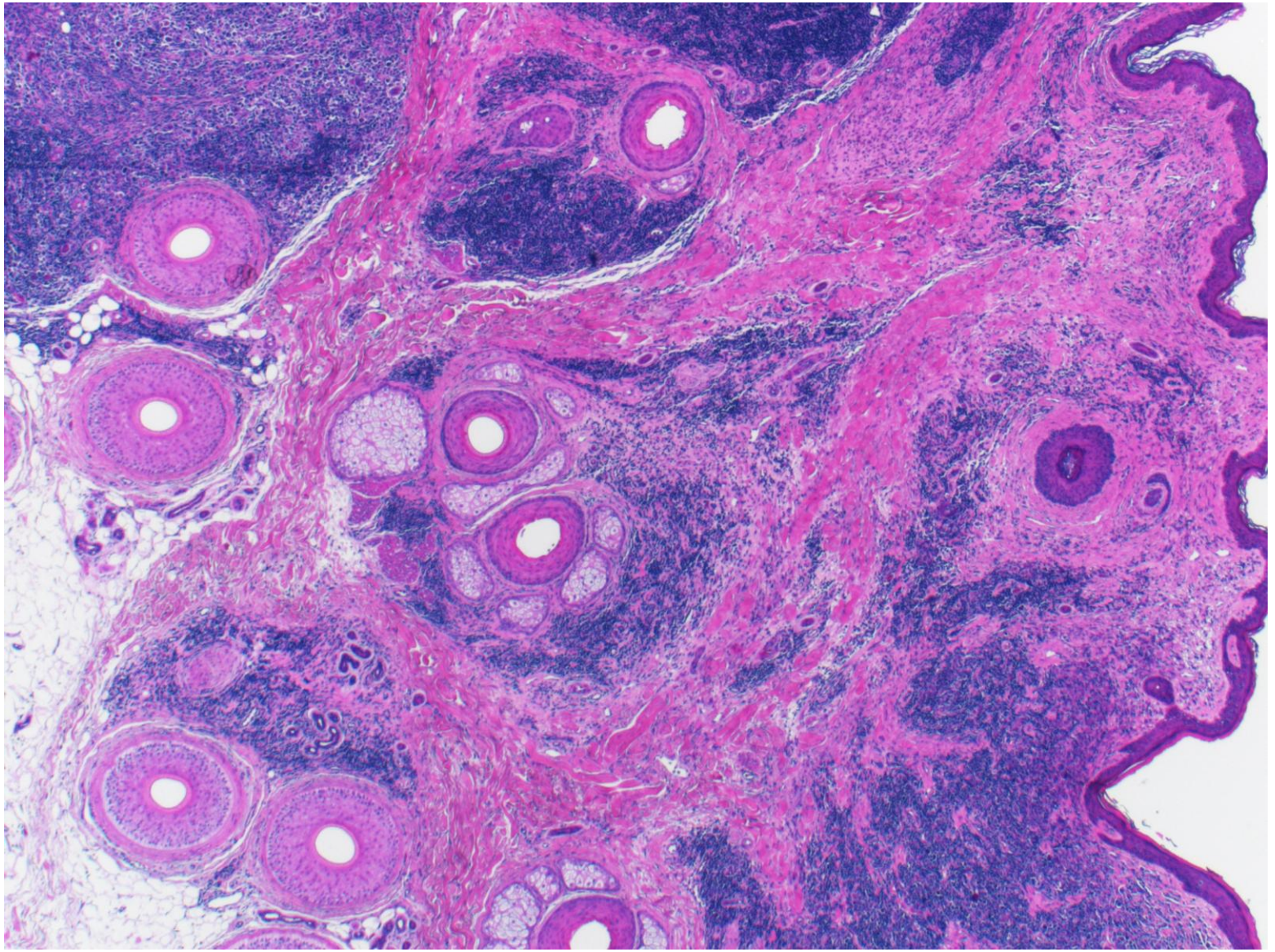
Langerhans cell sarcoma

According to the classification by the WHO, dendritic cell neoplasms include Langerhans cell histiocytosis, Langerhans cell sarcoma, interdigitating dendritic cell sarcoma/tumor, follicular dendritic cell sarcoma/tumor and dendritic cell sarcoma. Langerhans cell sarcoma should be distinguished from Langerhans cell histiocytosis, and it occurs most commonly in the skin and lymph node, but may be seen in the lung, liver, spleen and bone marrow. The skin lesions most commonly involve the trunk, scalp and legs. The prognosis is poor, particularly in cases with multi-organ involvement. Microscopically, the malignant Langerhans cells accompany nuclear atypia with pleomorphism and increased mitoses, The presence of longitudinal grooves in the nuclei is characteristic. The tumor cells are immunoreactive for S-100 protein, CD1a and CD207 (Langerin). CD68, CD21, CD35, lysozyme, HLA-DR, CD4, fascin, factor XIIIa and cyclin D1 may also be expressed. Ultrastructurally, Birbeck granules can be detected.

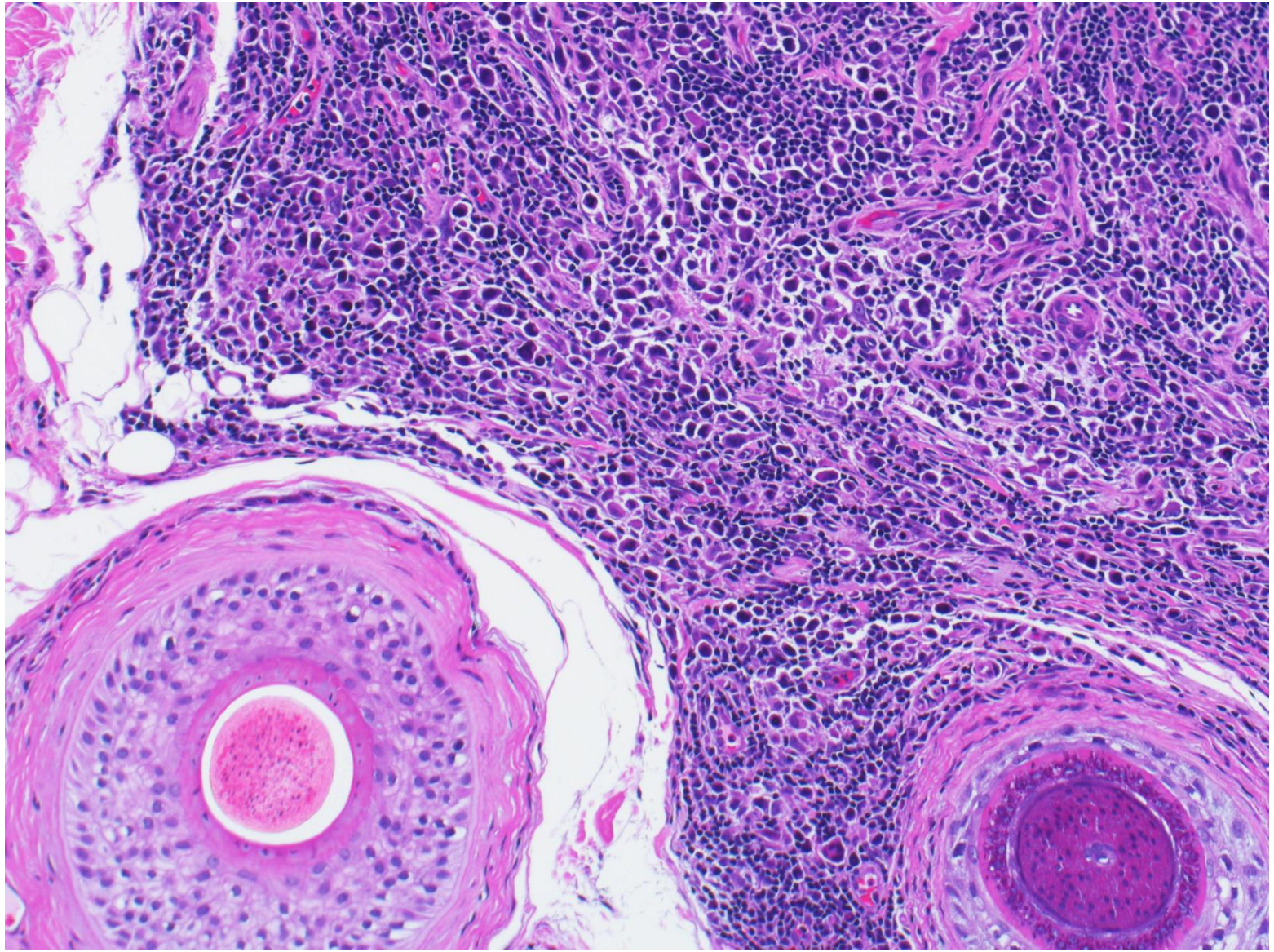
Ref.: Howard JE, et al. Langerhans cell sarcoma: a systematic review. Cancer Treat Rev 2015; 41(4): 320-331. doi: 10.1016/j.ctrv.2015.02.011



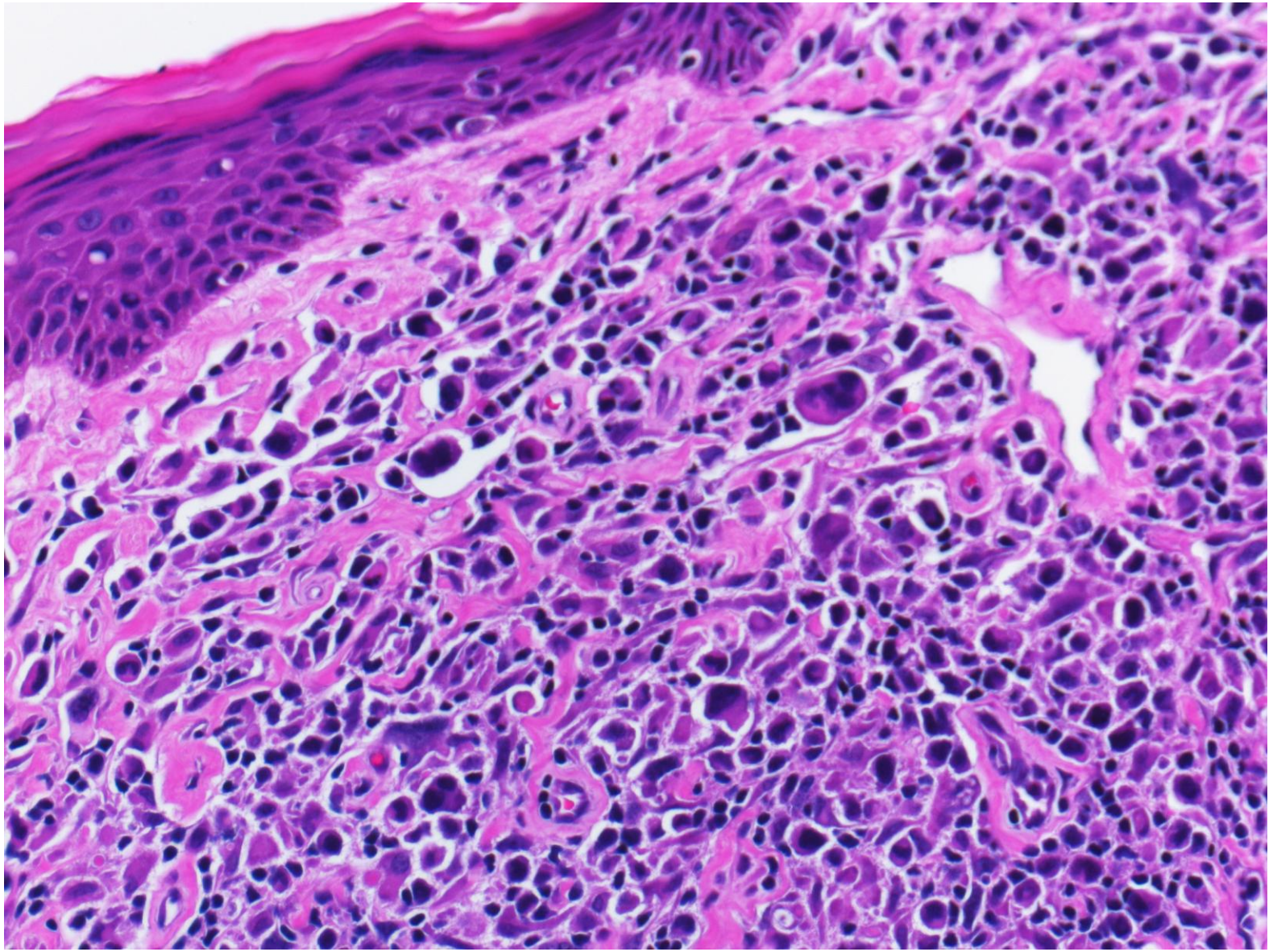
Recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. He received tumorectomy 3 months earlier. Hyperchromatic rounded tumor cells cluster around the skin appendage in the dermis (H&E-1).



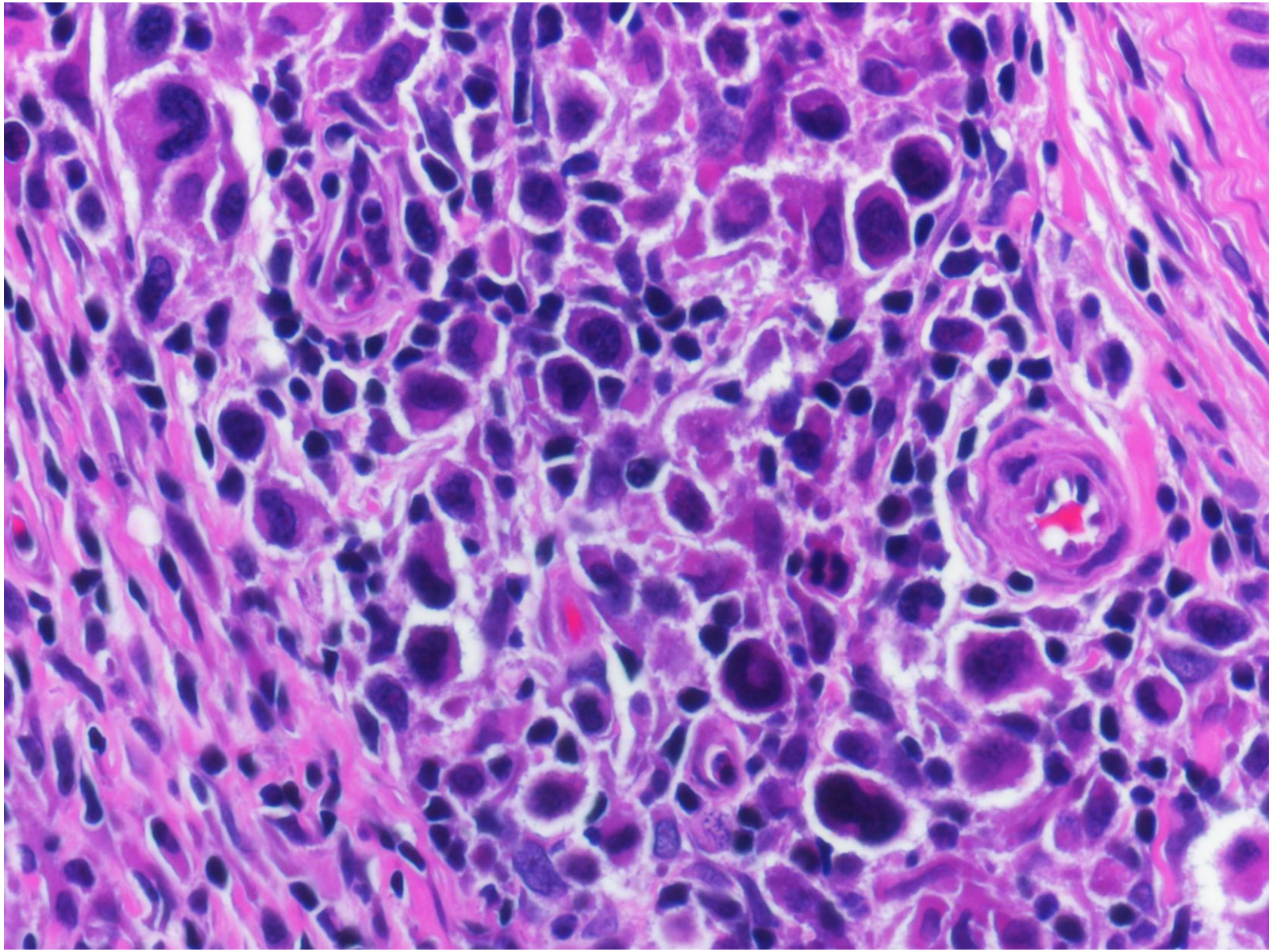
Recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. He received tumorectomy 3 months earlier. Hyperchromatic rounded tumor cells cluster around the skin appendage in the dermis (H&E-2).



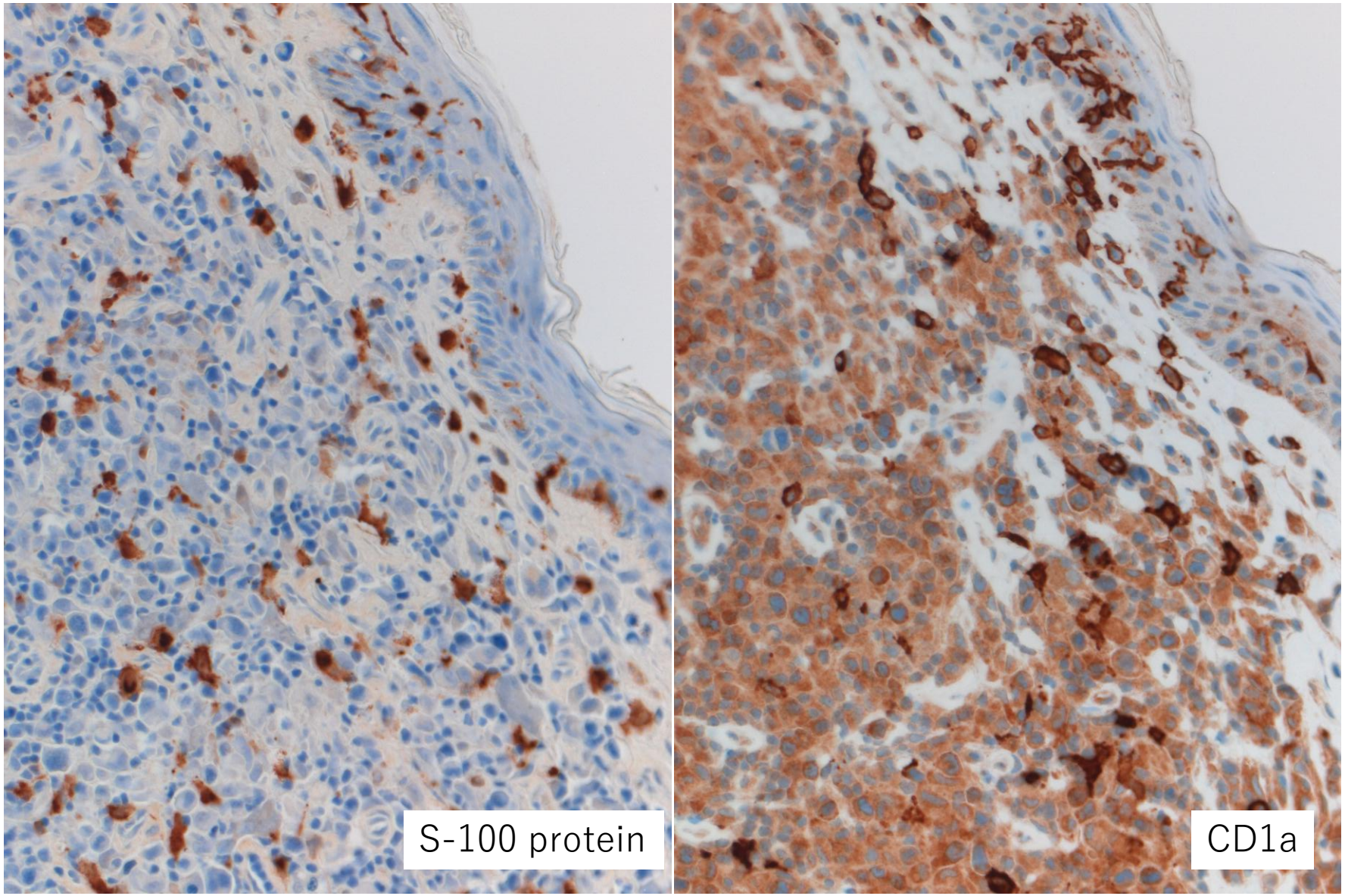
Recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. Hyperchromatic, poorly cohesive rounded tumor cells clustering around the skin appendage show nuclear pleomorphism (H&E-3).



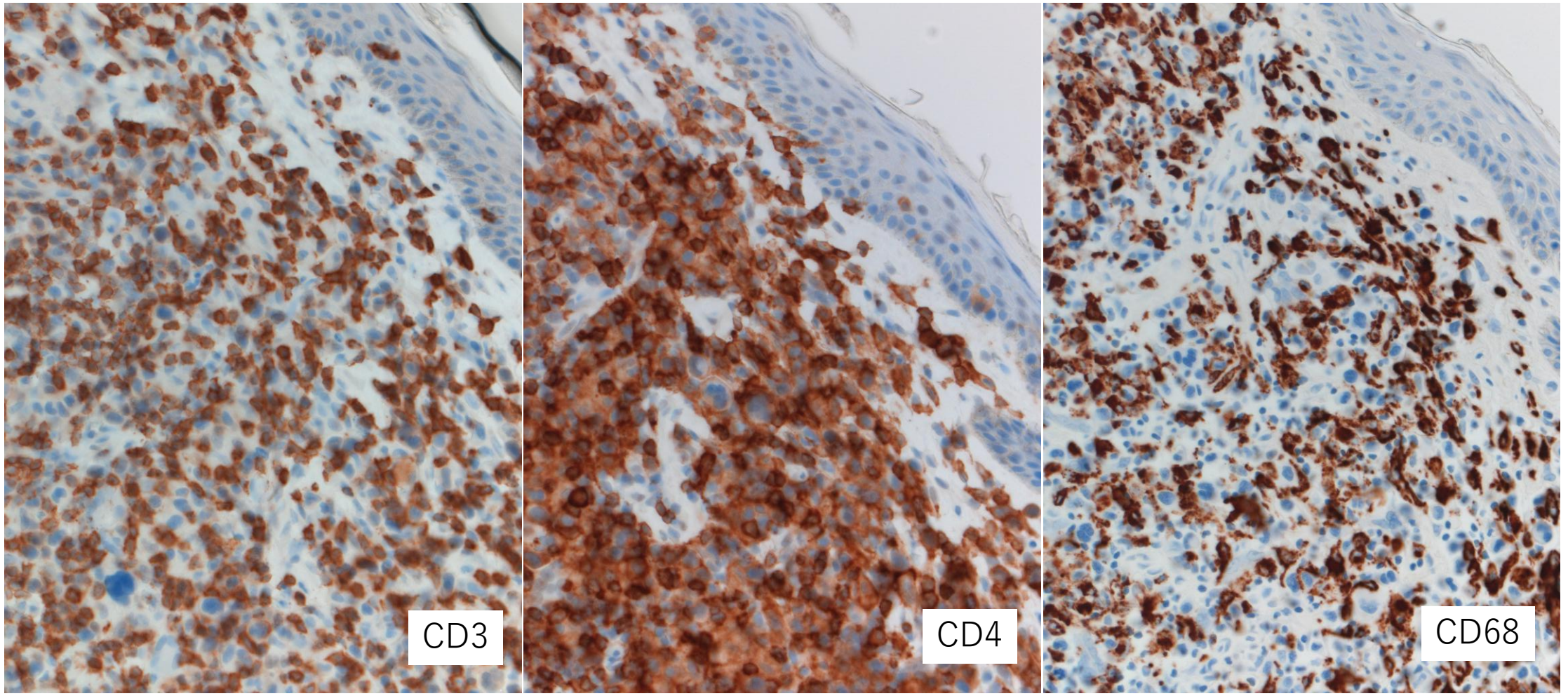
Recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. Hyperchromatic, poorly cohesive rounded tumor cells invading the dermis show nuclear pleomorphism (H&E-4).



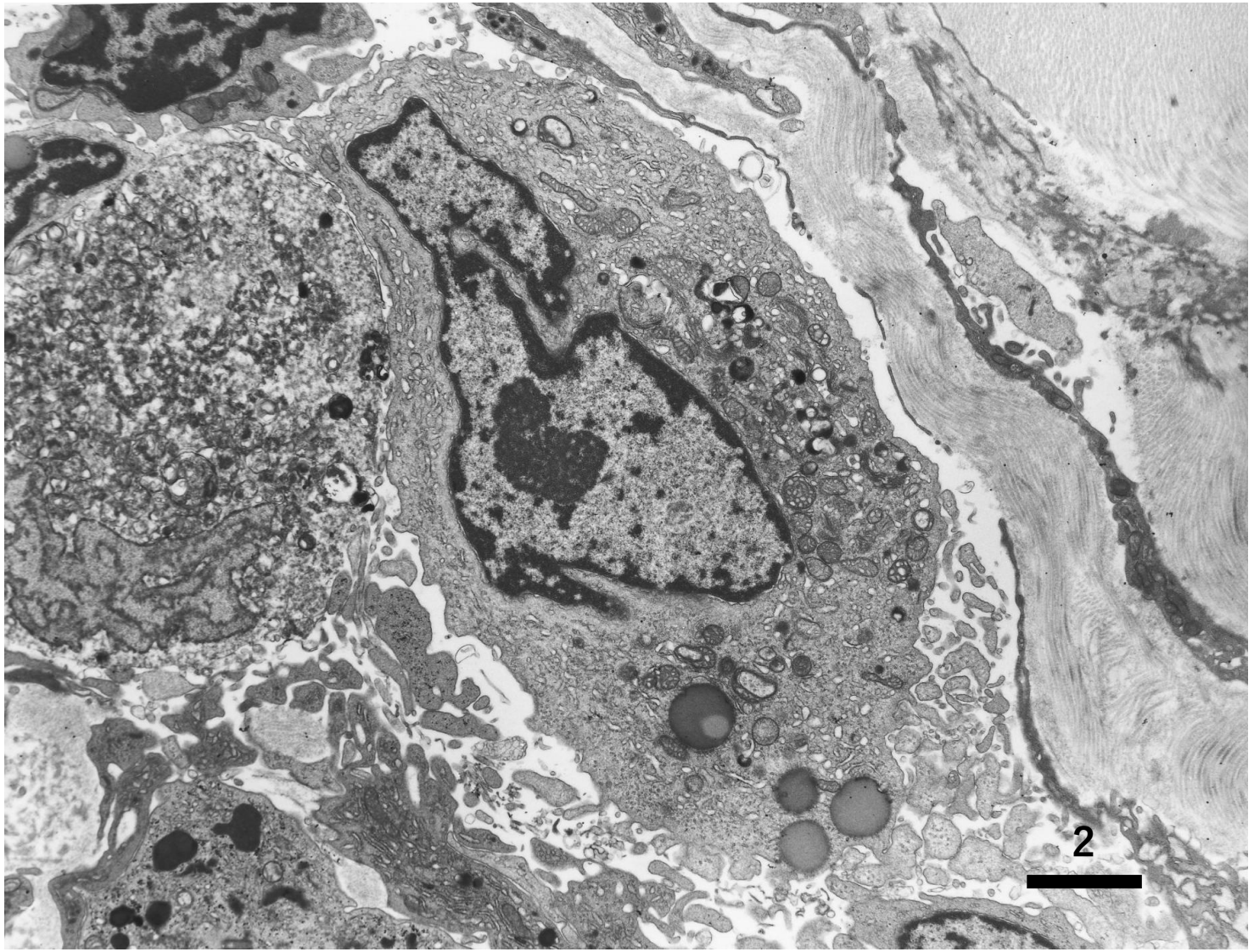
Recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. Hyperchromatic, poorly cohesive rounded tumor cells invading the dermis show nuclear pleomorphism. The nuclei are indented (H&E-5).



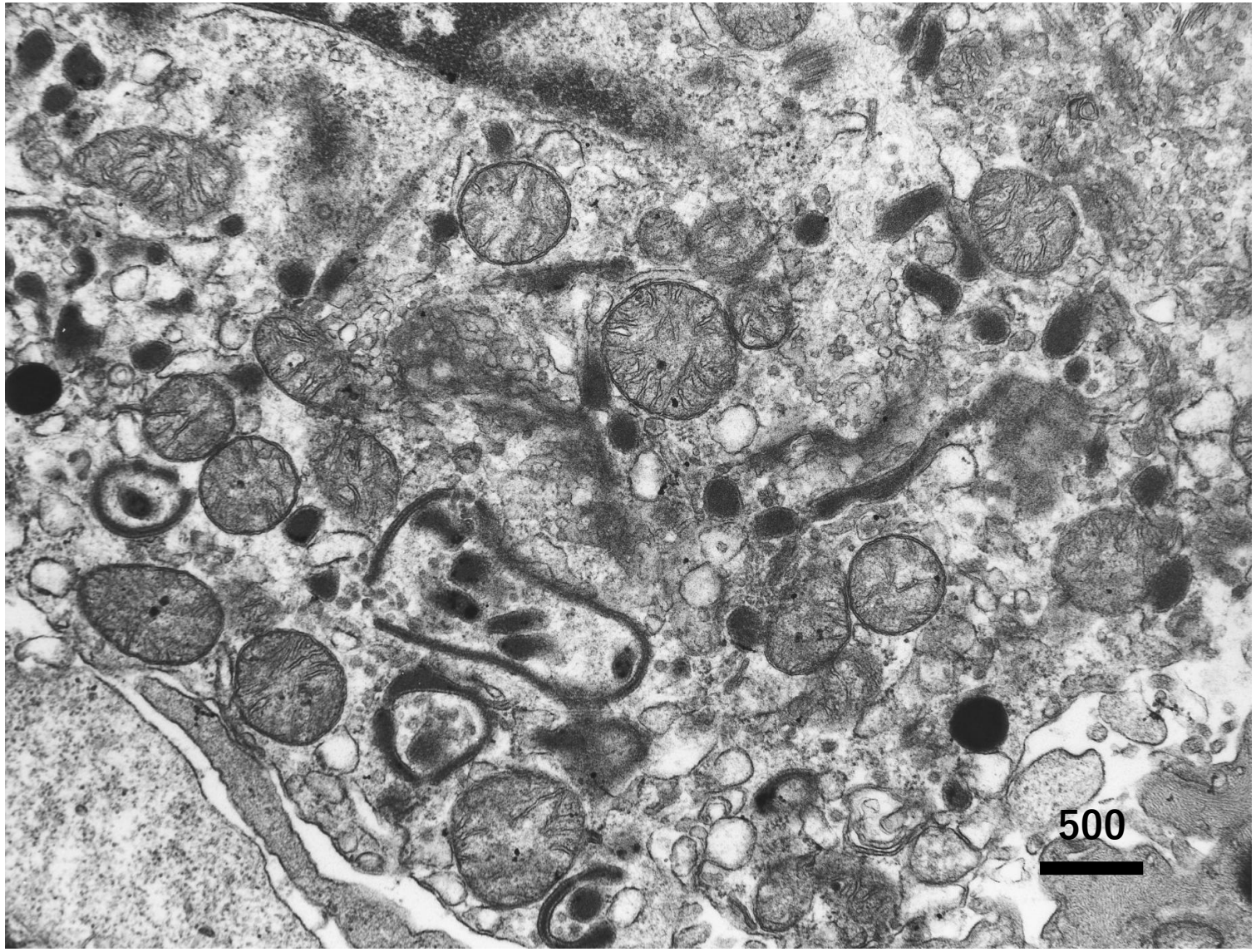
Recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. The tumor cells invading the dermis express S-100 protein and CD1a, confirming Langerhans cell origin. Intraepidermal Langerhans cells are increased (immunostaining for S-100 protein and CD1a).



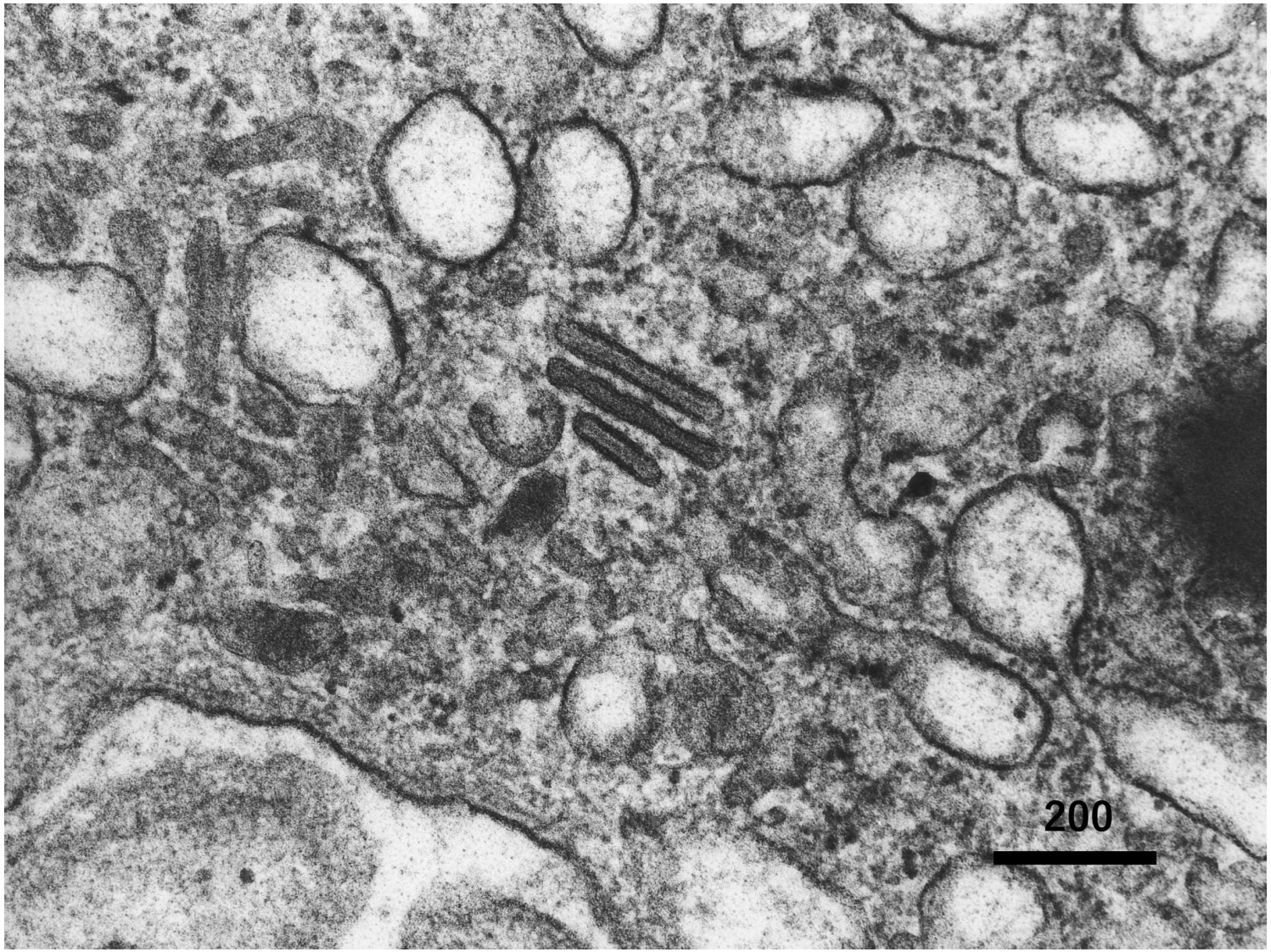
Recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. The tumor cells invading the dermis express CD3, CD4 and CD68. CD21 is not expressed in the present case (immunostaining for CD3, CD4 and CD68).



Ultrastructure of recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. The tumor cell possesses indented nucleus, prominent nucleolus and coarse lysosomal granules. Cytoplasmic podia formation is noted (EM-1).



Ultrastructure of recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. The tumor cell possesses electron-dense elongated bodies in the cytoplasm (being consistent with Birbeck granules) (EM-2).



Ultrastructure of recurrent Langerhans cell sarcoma of the ear lobe seen in an 82 y-o male patient. The tumor cell possesses Birbeck granules (elongated bodies with a central linear density and a striated appearance) in the cytoplasm (EM-3).