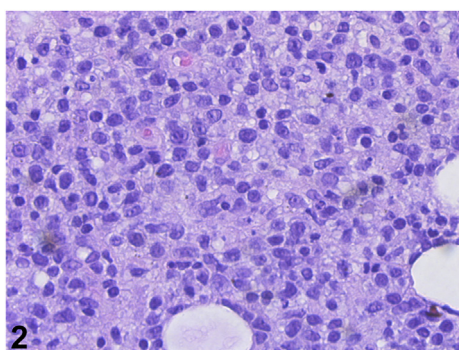


Erythematous, telangiectatic nodule within the melomental fold



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Key words: anaplastic large cell lymphoma; cutaneous T-cell lymphoma; dermatopathology.



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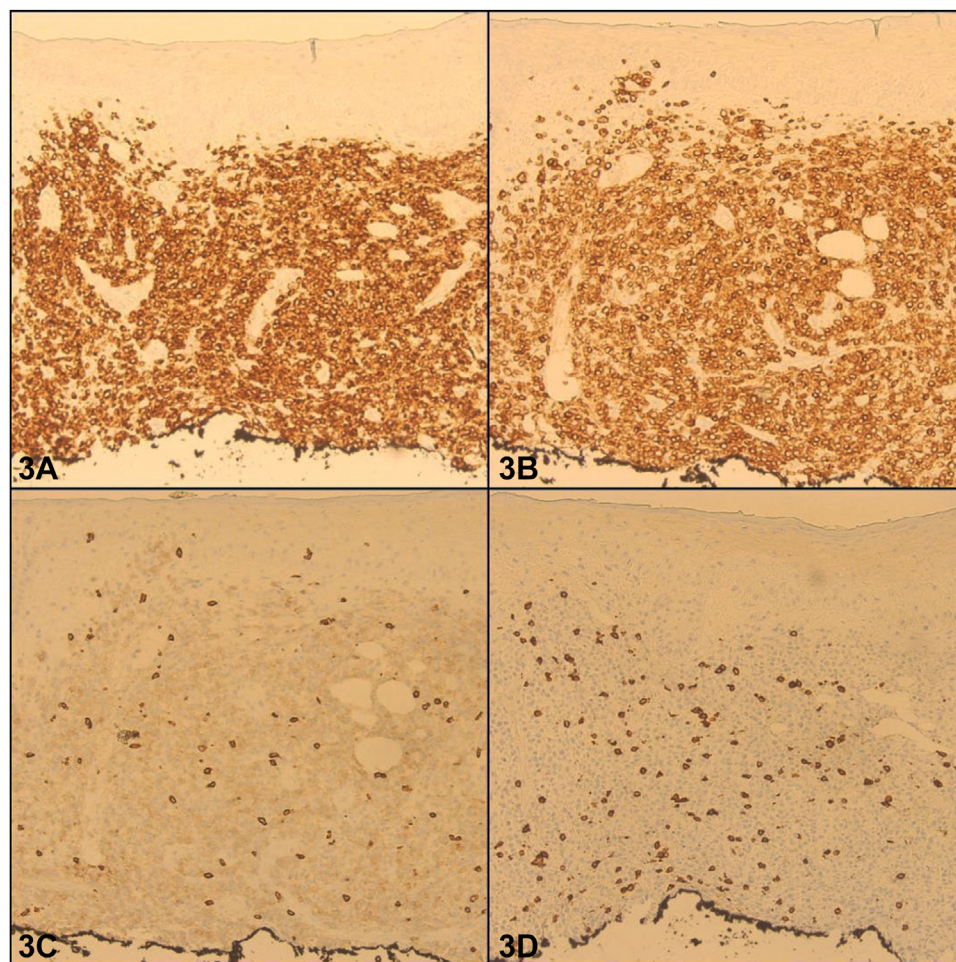
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A 79-year-old Caucasian man with a history of lymphoma presented with a 4-month history of an enlarging lesion of his left melomental fold. Other than occasional hemorrhage, the lesion was asymptomatic. Physical examination demonstrated a 2.5-cm erythematous, telangiectatic nodule (Fig 1). A deep shave biopsy was performed for histopathologic evaluation, revealing a nodular and diffuse dermal infiltrate of atypical medium-to-large lymphoid cells (Fig 2). Immunohistochemical studies demonstrated that the lymphoid infiltrate was CD30+, CD3+, CD8-, and CD20- (Fig 3). There was loss of CD5 expression, and CD4 was weakly expressed.

Question 1: What is the most likely diagnosis of the lesion of concern based on the information provided above?

- A. Mycosis fungoides, tumor stage
- B. Merkel cell carcinoma
- C. Kaposi sarcoma, nodular stage
- D. Cutaneous anaplastic large cell lymphoma
- E. Hodgkin lymphoma

Answers:

A. Mycosis fungoides, tumor stage — Incorrect. Although a biopsy of tumor-stage mycosis

fungoides may demonstrate a similar staining pattern, it is unlikely that this entity would be present as a solitary tumor as in this case. Patches and plaques would likely also have been observed.¹

B. Merkel cell carcinoma — Incorrect. The pathologic cells of Merkel cell carcinoma are CD3- and CD30-. This tumor exhibits a characteristic perinuclear dot staining pattern of CK20.¹

C. Kaposi sarcoma, nodular stage — Incorrect. Clinically, nodular-stage Kaposi sarcoma can present similarly to the lesion of concern in this patient. Histopathologic evaluation of nodular-stage Kaposi sarcoma, however, would demonstrate spindled and epithelioid endothelial cells along with

characteristic slit-like vascular spaces forming a “sieve-like” pattern. Positive immunohistochemistry staining of latency-associated nuclear antigen of human herpesvirus 8 is both sensitive and specific for Kaposi sarcoma.¹

D. Cutaneous anaplastic large cell lymphoma (ALCL) – Correct. The above histologic phenotype, along with the patient’s clinical presentation, is consistent with a diagnosis of cutaneous ALCL. On histology, the lymphoid cells in ALCL classically demonstrate round, oval, or irregular nuclei; striking nucleoli, and ample cytoplasm. Additionally, the cutaneous form of ALCL is typically anaplastic lymphoma kinase-negative compared with the systemic form of the disease. Anaplastic lymphoma kinase positivity may suggest a t(2;5) translocation.¹ This patient had a known history of cutaneous ALCL.

E. Hodgkin lymphoma – Incorrect. Hodgkin lymphoma is a B-cell neoplasm that rarely involves the skin; thus, expression of CD3 is unlikely. Additionally, the malignant cells of lymphocyte-dominant Hodgkin lymphoma are CD20+.¹

Question 2: Which of the following features are most likely observed based on pathologic evaluation of this entity?

- A. Bean bag cells
- B. Reed-Sternberg cells
- C. Hallmark (horseshoe) cells
- D. Clover leaf (flower) cells
- E. Dutcher bodies

Answers:

A. Bean bag cells – Incorrect. “Bean bag cells” describe cytophagic histiocytes. These cells are observed in various lymphomas, including subcutaneous panniculitis-like T-cell lymphoma.¹

B. Reed-Sternberg cells – Incorrect. Reed-Sternberg cells are observed in Hodgkin lymphoma, and 30%-40% of patients with the classical form of this disease will have Epstein-Barr virus DNA within these cells. Reed-Sternberg cells are typically of B-cell origin in classic Hodgkin lymphoma.¹

C. Hallmark (horseshoe) cells – Correct. Some of the anaplastic cells observed in cutaneous ALCL can be described as having horseshoe-shaped nuclei and are referred to as “hallmark cells.”² In a minority of the cases, the lymphoid cells of ALCL have

been described as pleomorphic, immunoblastic, or Reed-Sternberg-like.¹

D. Clover leaf (flower) cells – Incorrect. Clover leaf (flower) cells are large, atypical lymphocytes observed in adult T-cell leukemia/lymphoma. The cells are named as such because of the irregularity of their nuclear contour.³

E. Dutcher bodies – Incorrect. Dutcher bodies are associated with some B-cell neoplasms, such as primary cutaneous marginal-zone B-cell lymphoma and appear as an eosinophilic, Periodic acid-Schiff–positive inclusion within the nucleus. They are thought to represent immunoglobulins or glycoproteins.¹

Question 3: Which of the implantable devices listed below has been associated with an increased risk developing the noncutaneous form of this patient’s disease?

- A. Implantable cardioverter-defibrillator
- B. Breast implant
- C. Synthetic vascular graft
- D. Long-term venous access device
- E. Prosthetic heart valve

Answers:

A. Implantable cardioverter-defibrillator – Incorrect. To our knowledge, there have been no reported cases of ALCL associated with this type of device.⁴

B. Breast implant – Correct. A study by de Boer et al found that breast implants were associated with an elevated relative risk of 421.8 of breast ALCL. All of the breast implants found to be associated with breast ALCL in the study were permanent, ipsilateral, and silicone-filled. Out of the 28 cases where the type of breast implant was known, 23 were macrot textured. None were smooth or polyurethane-coated.⁵

C. Synthetic vascular graft – Incorrect. To our knowledge, synthetic vascular grafts have not been reported to be associated with cutaneous ALCL.⁴

D. Long-term venous access device – Incorrect. To our knowledge, long-term venous access devices have not been reported to be associated with cutaneous ALCL.⁴

E. Prosthetic heart valve – Incorrect. To our knowledge, prosthetic heart valves have not been reported to be associated with cutaneous ALCL.⁴

Abbreviation used:

ALCL: anaplastic large cell lymphoma

Conflicts of interest

None disclosed.

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