Solitary pink papule in an elderly man

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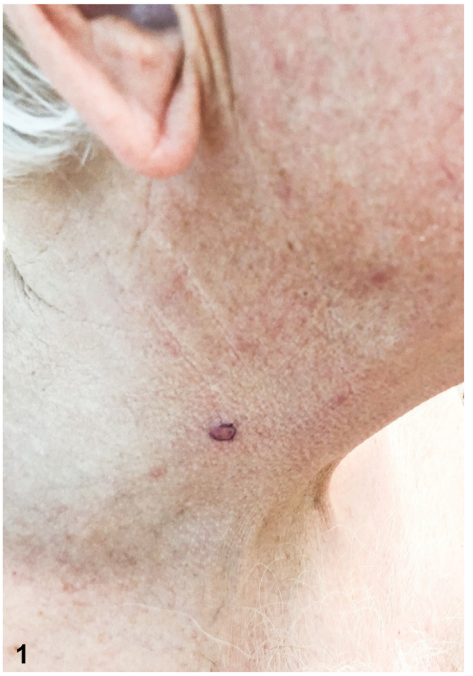
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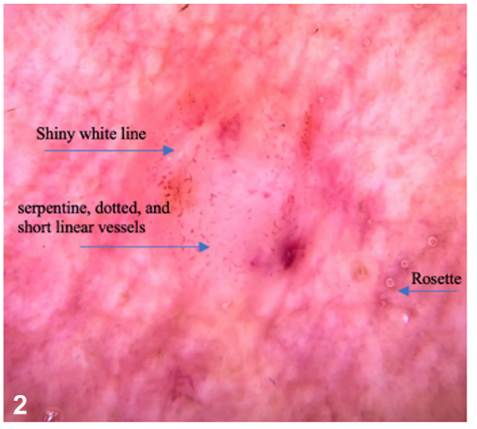
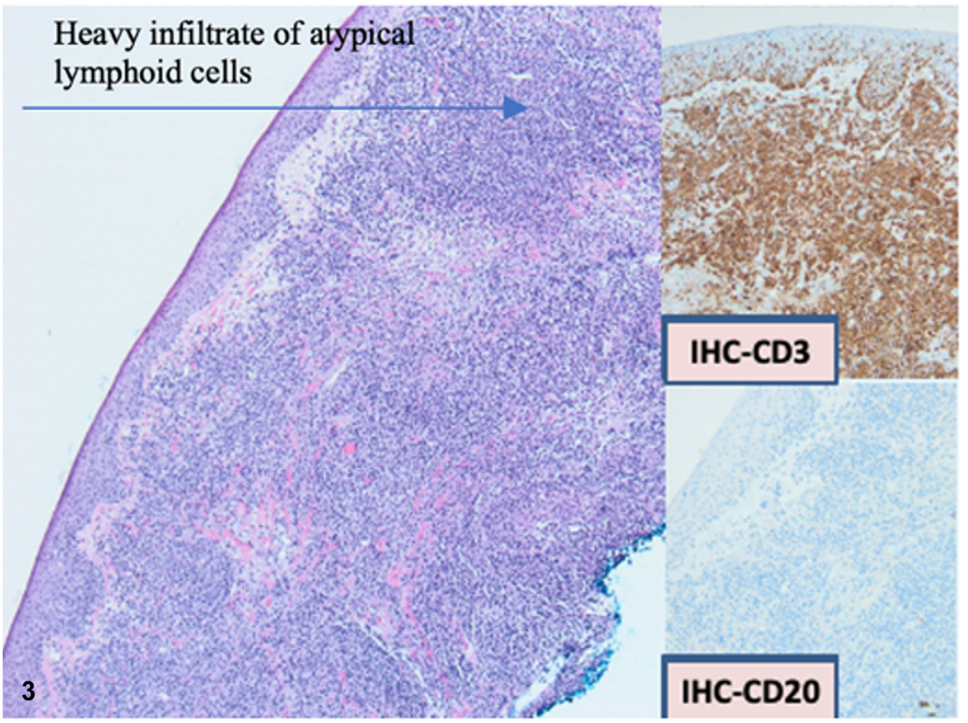
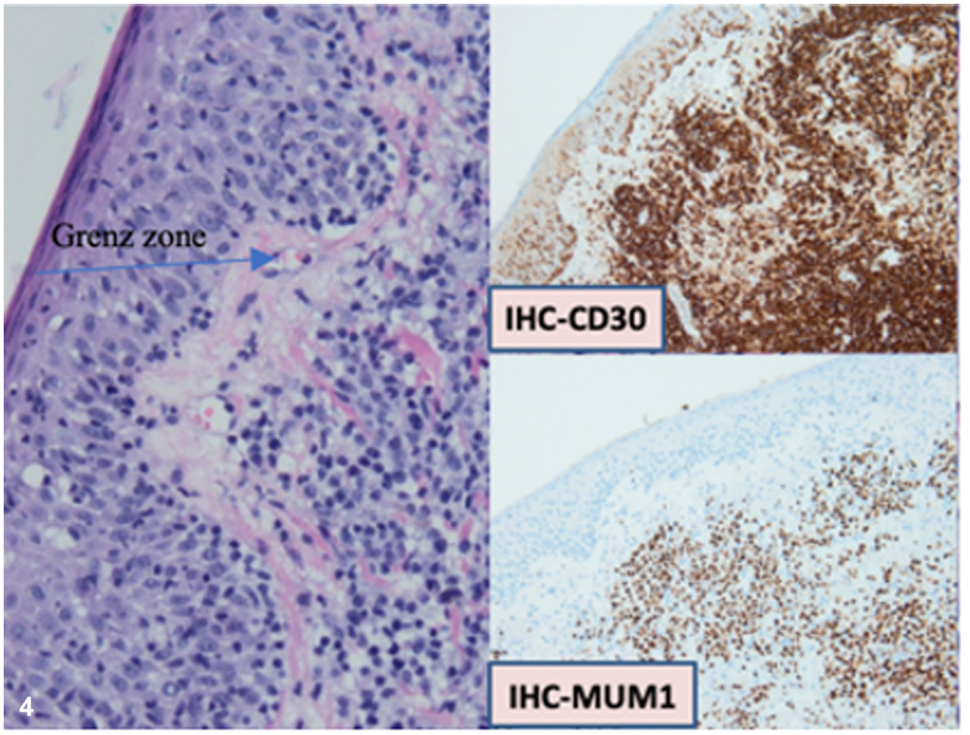



Heavy infiltrate of atypical lymphoid cells
IHC-CD3
IHC-CD20
3


Grenz zone
IHC-CD30
IHC-MUM1
4

A 72-year-old, healthy man presented with an asymptomatic, stable, solitary, 5-mm × 5-mm, pink, dome-shaped papule on his neck that had been present for at least 6 months (Fig 1). There were no identifiable triggers, including exposure to drugs, radiation, or trauma to that area. A physical examination was unremarkable and did not demonstrate lymphadenopathy or hepatosplenomegaly. Polarized dermoscopy demonstrated a homogeneous, pink papule with serpentine, dotted, and short linear vessels; white, shiny lines; and white rosette structures (Fig 2). Pathology showed a heavy infiltrate of intermediate-to-large, atypical lymphoid cells involving the papillary dermis, dermoepidermal junction, and epidermis. The atypical lymphoid cells monotonously stained for CD3, diffusely expressed CD30 and nuclear MUM1 (bright), and were double-negative for CD4 and CD8 (Figs 3 and 4). ALK-1, SOX10, PD1, and CD138 were negative. No further investigations were undertaken.

Question 1: Given the history and images, what is the most likely diagnosis?

A. Primary cutaneous anaplastic large cell lymphoma with 6p25.3 rearrangement

B. Transformed $CD30^+$ mycosis fungoides (MF)

C. Lymphomatoid papulosis (LyP) with 6p25.3 rearrangement (LyP 6p25.3)

D. LyP type C

E. LyP type B

Answers:

A. Primary cutaneous anaplastic large cell lymphoma with 6p25.3 rearrangement — Incorrect. Histologically, this entity can also present with a nodular dermal infiltrate of $CD30^+$, atypical lymphoid cells but no evidence of epidermal involvement of $CD30^+$ cells, which is one of characteristic features of this subtype of LyP.[1,2] The 6p25.3 rearrangement on the *DUSP22* locus has been correlated with the inhibition of tumor suppression.[3]

B. Transformed $CD30^+$ MF — Incorrect. This patient had no prior diagnosis of MF, and MF does not usually present with one solitary papule on the head and neck region.

C. LyP 6p25.3 — Correct. This entity was first reported by Karai et al[4] in 2013. It usually presents in men, at a mean age of 75, as one or a few papulonodules at a single site. Pathology reveals a dermal nodule composed of lymphocytes with overlying $CD30^+$, $MUM1^+$, atypical lymphocytes in the epidermis, resembling pagetoid reticulosis.[2,4] In general, surveillance is recommended for patients with LyP given the risk for developing secondary lymphomas; however, Karai et al[4] did not note the development of lymphomas in any of their patients.

D. LyP type C — Incorrect. LyP type C shares certain histologic features of LyP with 6p25.3 rearrangement, such as nodular infiltrates in lymphocytes; however, CD4 and TIA1 are usually also expressed.[2,4] The epidermal involvement of $CD30^+$ lymphoid cells is not a characteristic feature of LyP type C. Clinically, it presents at a mean age of 38 and is typically characterized by disseminated papulonodules.[4,5]

E. LyP type B — Incorrect. The epidermal findings of LyP 6p25.3 are similar to those of LyP type B; however, LyP type B expresses CD4 and has more of a band-like dermal lymphocytic infiltrate with no evidence of the epidermal involvement of $CD30^+$ lymphoid cells.[2,4,5]

Question 2: What is the common immunohistochemical staining pattern found in LyP with a 6p25.3 rearrangement?

A. $CD3^+$, $CD30^+$, $MUM1^+$, ALK^-, CD4, and CD8 are often (but not always) negative

B. $CD30^+$, $CD4^+$, ALK^-, EMA^-, CLA^+

C. $CD4^+$, $CD8^-$, $CD30^{+/-}$

D. $CD4^+$, $CD8^-$, $CD30^+$

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E. $CD4^+$ greater than $CD8^+$, $CD30^{+/-}$, $CD3^-$, $CD7^-$

Answers:

A. $CD3^+$, $CD30^+$, $MUM1^+$, ALK^-, CD4, and CD8 are often (but not always) negative — Correct. A review of 11 patients with LyP 6p25.3 showed this pattern most often.[4] Of note, the CD30 stain is more prominent in the dermis than in the epidermis in most reported cases; this was seen in our patient as well (Fig 4).[2,4]

B. $CD30^+$, $CD4^+$, ALK^-, EMA^-, CLA^+ — Incorrect. This is the staining pattern for primary cutaneous anaplastic large cell lymphoma.[6]

C. $CD4^+$, $CD8^-$, $CD30^{+/-}$ — Incorrect. This is the staining pattern for LyP type B.[6]

D. $CD4^+$, $CD8^-$, $CD30^+$ — Incorrect. This is the staining pattern for LyP type C.[6]

E. $CD4^+$ greater than $CD8^+$, $CD30^{+/-}$, $CD3^-$, $CD7^-$ — Incorrect. This is the staining pattern for transformed MF.

Question 3: Which of the following is a dermoscopic pattern for LyP 6p25.3?

A. A white, structureless area surrounding tortuous vessels that extends centrifugally

B. A central, white, structureless area with vessels present on the periphery

C. Vessels surrounding an area of necrosis and ulceration

D. A brown-gray, structureless area with no vessels

E. Linear, curved, and tortuous vessels; rosettes; and shiny, white lines

Answers:

A. A white, structureless area surrounding tortuous vessels that extends centrifugally — Incorrect. This pattern is seen in the initial stage of more common forms of LyP.[5,7]

B. A central, white, structureless area with vessels present on the periphery — Incorrect. This pattern is seen with more mature lesions of more common forms of LyP.[5,7]

C. Vessels surrounding an area of necrosis and ulceration — Incorrect. This pattern is seen in more common forms of LyP prior to the cicatricial phase.[5,7]

D. A brown-gray, structureless area with no vessels — Incorrect. This pattern is seen with scarring or postinflammatory dyspigmentation in more common forms of LyP.[5,7]

E. Linear, curved, and tortuous vessels; rosettes; and shiny, white lines — Correct. To our knowledge, this pattern has not yet been described, and it was seen in our patient with LyP with 6p25.3 gene rearrangement.

Abbreviations used:

LyP: lymphomatoid papulosis
LyP 6p25.3: lymphomatoid papulosis with 6p25.3 rearrangement
MF: mycosis fungoides

Conflicts of interest

None disclosed.

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