2018 WHO-EORTC classification update

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Willemze R, Cerroni L, Kempf W, Berti E, Facchetti F, Swerdlow SH, Jaffe ES. The 2018 update of the WHO-EORTC classification for primary cutaneous lymphomas. *Blood*. 2019;133(16):1703-1714.

1. Your patient is suspected of having a primary cutaneous lymphoma. According to the 2018 update of the World Health Organization–European Organization for Research and Treatment of Cancer (WHO-EORTC) consensus classification by Willemze et al, which of the following statements about new provisional entities now included in the classification of primary cutaneous lymphomas is correct?

- ☐ Primary cutaneous acral $CD8^+$ T-cell lymphoma is a newly described entity that typically presents as a diffuse rash on the trunk
- ☐ Primary cutaneous acral $CD8^+$ T-cell lymphoma is highly aggressive and requires systemic chemotherapy
- ☐ An Epstein-Barr virus (EBV)–positive mucocutaneous ulcer (EBVMCU) is a newly described entity occurring primarily in immunocompetent patients
- ☐ EBVMCU lesions contain large Hodgkin-like EBV^+ B cells ($PAX5^+$) in a mixed inflammatory background with variable CD20 expression and a non–germinal center phenotype

2. According to the 2018 update of the WHO-EORTC classification by Willemze et al, which of the following statements about modifications in the sections on lymphomatoid papulosis (LyP) and primary cutaneous marginal zone lymphoma (PCMZL) is correct?

- ☐ The 2018 update recognizes only 3 subtypes of LyP
- ☐ A newly described LyP subtype has chromosomal rearrangements involving the *DUSP22-IRF4* locus on chromosome 6p25.3
- ☐ The 2018 update recognizes 4 different subtypes of PCMZL
- ☐ The most common PCMZL subtype contains neoplastic B cells that express immunoglobulin M and often the chemokine receptor CXCR3

3. According to the 2018 update of the WHO-EORTC consensus classification by Willemze et al, which of the following statements about other changes in the classification of primary cutaneous lymphomas is correct?

- ☐ A new section describes cutaneous forms of chronic active EBV disease in childhood, which includes hydroa vacciniforme–like lymphoproliferative disease and hypersensitivity reactions to mosquito bites
- ☐ The terminology and definitions of primary cutaneous lymphoma types in the 2018 update differ markedly from those in the 2017 *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues* blue book
- ☐ Primary cutaneous $CD4^+$ small/medium T-cell lymphoproliferative disorder is now known as "primary cutaneous $CD4^+$ small/medium T-cell lymphoma"
- ☐ Genetic markers have replaced histologic, immunophenotypic, genetic, and clinical data for accurate diagnosis

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