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EBV$^+$ mucocutaneous ulcer: a new entity of WHO 2017

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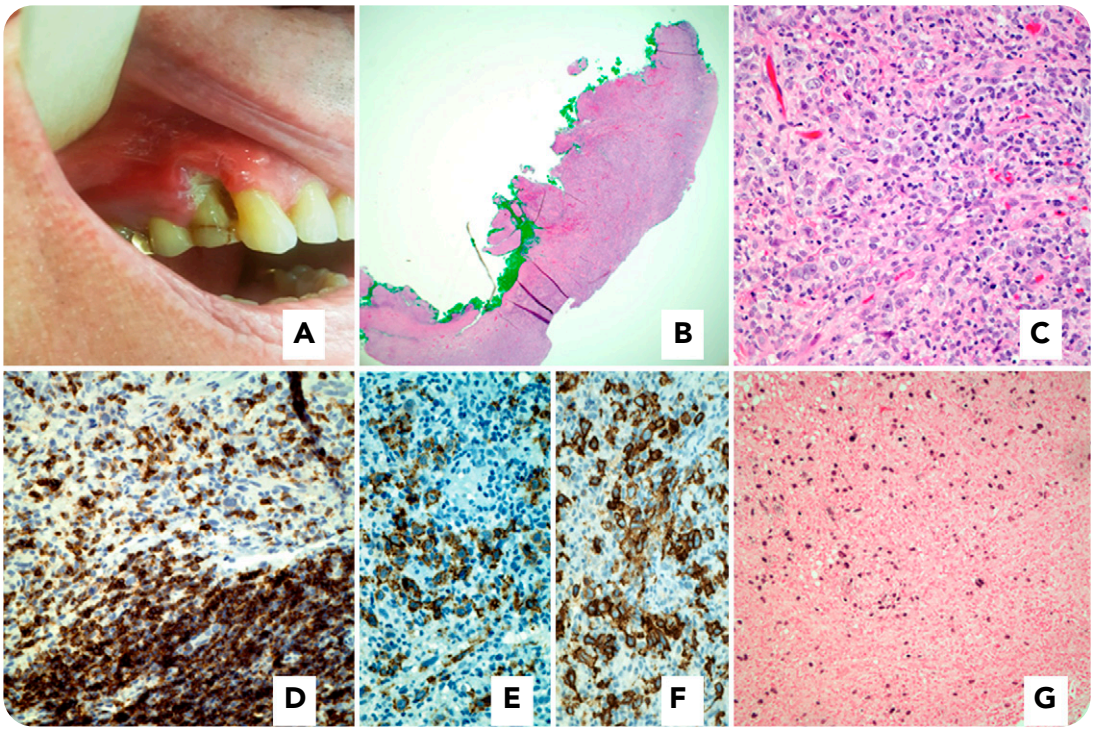

A 79-year-old apparently healthy man presented with an isolated, persistent oral lesion. On examination, a single, well-circumscribed ulcerative gingival lesion was identified measuring ~1 cm (panel A). No systemic lymphadenopathy or hepatosplenomegaly was identified. A biopsy demonstrated oral mucosa with surface ulceration and an underlying polymorphic, mixed inflammatory infiltrate, including clusters of large atypical lymphoid cells with somewhat Hodgkin-like morphology (panels B-C; hematoxylin and eosin stain, original magnification ×10 [B], ×40 [C]). Patchy areas of necrosis and focal angioinvasion were also identified. The atypical cells were positive for CD20 (panel E; immunohistochemistry stain, original magnification ×40), CD30 (panel F; immunohistochemistry stain, original magnification ×40), Epstein-Barr virus (EBV; panel G; EBV encoding RNA in situ hybridization, original magnification ×20), MUM1, PAX-5, and BCL-6, but negative for CD10, CD45, and ALK-1 protein. On the peripheral edge of the lesion, there were dense small lymphocytic infiltrates, which were rich in CD3$^+$ T cells (panel D; immunohistochemistry stain, original magnification ×20). The clinical and histologic characteristics are typical for EBV$^+$ mucocutaneous ulcer (EBVMCU).

EBVMCU is a newly recognized clinicopathologic entity in the 2017 revision of World Health Organization (WHO) classification. The disease often occurs in elderly patients, sometimes in the setting of iatrogenic immunosuppression, which plays a role in pathogenesis. EBVMCU usually presents as an isolated ulcerative lesion, most commonly involving the oral mucosa but also appearing in skin or gastrointestinal tract. The disease typically follows an indolent clinical course.

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