Our Dermatology Online

Letter to the Editor

Epstein–Barr virus-positive mucocutaneous ulcer in a child, a rare immunoblastic B lymphoma

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Sir,

Epstein–Barr virus-positive mucocutaneous ulcer (EBV+, MCU) is a temporary entity, recently included in the 2016 World Health Organization (WHO) classification of haemopathies malignancies [1]. We report an interesting clinical case that highlights this new entity in a 13-year-old child.

A 13-year-old girl, with no history, presenting chronic mucocutaneous nasal ulcerations, the clinical examination finds, a girl in good general condition with centro-facial ulcerations, (Figs. 1 and 2) without associated lymphadenopathy and the rest of the somatic examination was no abnormality. The biological and radiological exploration did not show any specific anomaly, the biopsy objectifying a polymorphic dermal infiltrate with sternbergoide type B cells, EBV + expressing the antigens CD15, CD30, CD20, (Figs. 3 and 3) MUM1 with reactive T lymphocyte pax positive. The extension assessment did not show any anomaly. The immunoglobulin weight assay showed no immunodeficiency.

The diagnosis of EBV-positive skin and mucosal ulcer of the WHO 2016 classification ofhaemopathies was retained and a treatment with poly chemotherapy (Rituximab, Vincristine, Cyclophosphamide and Doxorubicin) was initiated with excellent progress and a decline of 11 months without recurrence (Fig. 5).

(EBV+, MCU) presents with a chronic ulcer of the oropharynx, skin, gastrointestinal tract or rarely the

Figure 1: Mutilating centrofacial ulcerations.

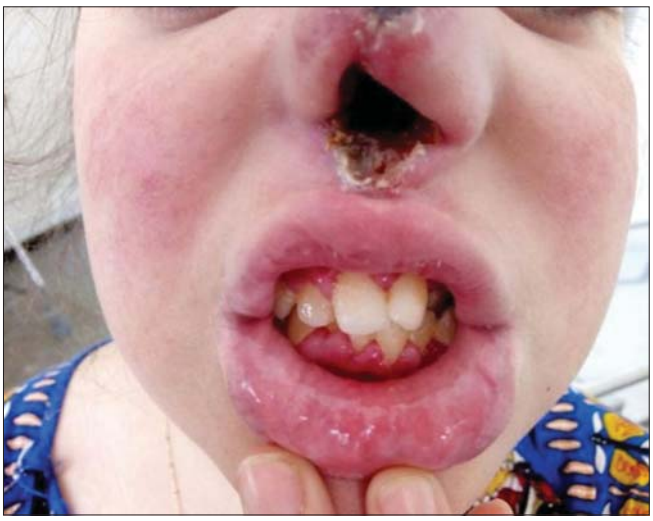

Figure 2: Destruction of the nasal septum.

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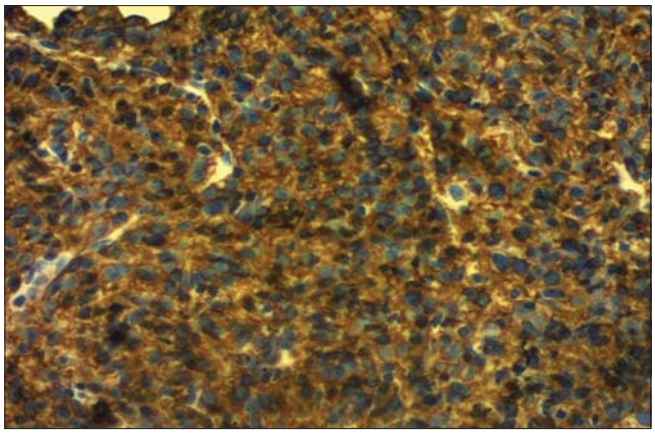

Figure 3: Diffuse labeling of lymphoma cells by CD20.

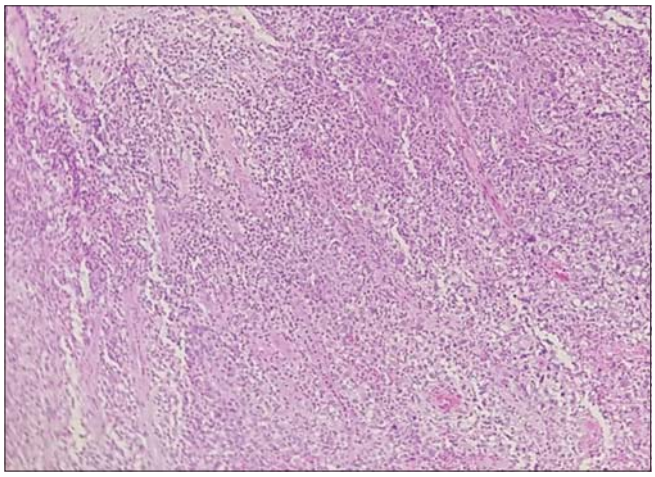

Figure 4: Biopsy of the nasal mucosa at low magnification.

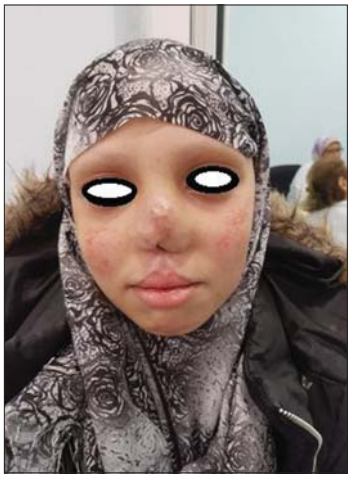

Figure 5: Improvement under chemotherapy.

genital mucosa. Histologically, (EBV+, MCU) is made of a polymorphic infiltrate with large sternbergoide B cells that express the EBV transcript, the CD15, CD30 and CD20 antigens, with reactive T cells [2]. The diagnosis with certainty is sometimes difficult, but generally the indolent clinical character without systemic involvement, with the possibility of spontaneous regression [2-5]. The presence of CD15 + and EBER + lymphoma cells without angiocentrism as well as the absence of B clonality are compatible with this diagnosis [6].

The treatment of this new entity is not yet well defined, the few cases reported in the literature have been treated either by standard chemotherapy, CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone), or by rituximab and bendamustine, others responded well to radiotherapy alone [7].

EBV+, MCU a difficult diagnostic entity, is important to identify the diagnostic criteria, the factors that determine its evolution and the need for treatment in order to provide an adequate diagnosis and better management.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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