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Primary cutaneous marginal zone b-cell lymphoma: Case report

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Abstract
Primary cutaneous marginal zone B-cell lymphoma is a very rare, indolent entity, classified in in the category of extranodal marginal zone B cell lymphoma, and which presents 2% of all cutaneous lymphomas. It is an entity with several clinical characteristics and evolutionary, and especially histological, sometimes common with other neighboring entities, and posing above all the problem of positive and differential diagnosis.
We expose a case of PCMZL with a particular presentation and difficulty of diagnosis over several years, insisting us through our observation on the necessity of repeating the anatomopathological examination with Immunohistochemistry
to be able to make the diagnosis. The treatment is not codified, the therapeutic modalities proposed by the experts are many and varied.

Keywords: marginal, Lymphoma, histological, entities

Introduction
Primary cutaneous marginal zone B-cell lymphoma (PCMZL) is a rare, indolent entity now classified in the category of extranodal marginal zone B cell lymphoma [1], We expose the case of a young patient presenting recurrent tumors, the diagnosis of PCMZL was retained after several biopsies and surgical excision.

Observation
a young patient of 24 years old, with no notable history, who presents for more than 8 years, subcutaneous tumors on the level of the 2 arms with very slow progression, and an autoregressive character, one of the tumors sits on the scar of an old vaccine. The initial biopsy was in favor of a nonspecific inflammatory infiltrate, but seen the progression, it was operated twice with recurrence of the tumor lesions, the anatomopathological readings revealed a pseudolymphoma B with possibly a Kimura disease,
on examination, in addition to the scars of the old surgical excision at the level of the right arm and the homolateral elbow, we objectified subcutaneous masses which are difficult to locate in relation to the underlying plane, poorly limited, painless, overcome with eczematiform reactions in the skin
The treatment initially chosen was a surgical excision, but the rereadings of the operating pieces revealed a PCMZL, after the final diagnosis, the tumors had regressed, the patient only kept a 1 cm subcutaneous nodule on the right arm; we opted before for an intralesional corticosteroid ingection of this lesion, with monitoring of the rest of the scars of the old lesions. The decline is underway
We highlight the difficulties encountered in the diagnosis of this particular type of cutaneous lymphoma.

Discussion
Primary cutaneous B-cell lymphoma (PCLB) is defined as a B-cell lymphoproliferative disorders limited to the skin and without any sign of extra-cutaneous disease at the time of diagnosis [1]. In the WHO classification, PCMZL are not recognized as a separate entity, they belong to the group of "MALT" lymphomas. Within the group of marginal zone lymphomas they are qualified as extra-ganglionic forms which can be developed in particular at the expense of the mucous membranes (MALT: Mucosa-Associated Lymphoid Tissue) or the skin (cutaneous MALT or SALT). (2,3) PCMZL is a particular entity, it presents 25% of LCB and 2 to 7% of all primary cutaneous lymphomas, which makes it a very uncommon neoplasm. [4]. The exact pathogenesis of PCMZL is not clear, the infectious hypothesis stems from the association reported with molecular and was proposed by analogy to gastric and adnexal lymphoma, and the association with Lyme disease (Borrelia burgdorferi) [7] It generally affects adults of all ages with a median age of 53 years, the literature notes a slight male predominance (sex ratio M / F 1.5 to 2), (4.5).
Clinically It appears in the form of single or few red-purple papules or tumor nodules, most often multifocal, often at the trunk and upper limbs, rarely head and neck, evolution is generally slow, rarely towards ulceration. [4, 6, 7]. In rare cases a spontaneous regression of the lesions development of anetodermia can be observed making the diagnosis difficult and delayed for several years. [8]. Another diagnostic difficulty can arise when lesions are made of papules and small nodules, without associated tumor lesions; Reminiscent of inflammatory or benign conditions such as lupus erythematosus timidus, Jessner-Kanof infiltrate or benign lymphoid hyperplasia. [9]. Skin lesions are often associated with preservation of general condition without systemic signs, [10].
The diagnosis is based on histology in the absence of extra-cutaneous signs, notably Lymph nod involvement,. Histologically, Two architectural forms are possible, either an

infiltrate mainly perivascular diffuse lymphoplasmacytic; either plurinodular infiltrate, especially periannexiliary, [4]. often comprising several cells, especially centrocytes from the marginal zone, abundant lymphoplasmocytoid cells, mixed with some centroblastic or immunoblastic type cells and reaction T cells often numerous.

Reactive lymphoid follicles are frequent, with a more or less marked hyperplasia of the marginal zone, and plasma cells arranged at the periphery (resembling a pseudolymphoma B, the case of our patient) [1, 11]. The epidermis is generally spared. Immunohistochemistry guides the final diagnosis: B cells in the area have a positive labeling with (CD20 +, CD79a +), bcl-2 +, but negative with CD10-, bcl-6-. (positive in the reaction follicles: CD10 +, Bcl6 +, Bcl2-), and CD5 -, (1, 7, 11, 12, 13]. Plasma cells express CD138 + but generally not CD20 (-), and detection of a monotype of the plasma cells at the periphery of the nodules, and this by calculating the ratio of the expression of the light chain of cytoplasmic immunoglobulins (lambda and kappa); It is a key element in the diagnosis, But it is inconsistent [4, 14]. Genetic studies report, in cases of PCMZL, standard translocations (14; 18) (q32; q21) involving the IGH gene on chromosome 14 and the MLT gene on chromosome 18 (found in MALTs of the eye appendices), and t (3; 14) (p14.1; q32) involving IGH and FOXP1 genes (found in extranodal MALTs) (1, 4, 7, 15, 16) But the type t (11; 18) (q21; q21) and t (1; 14) (p22; q32) transclocation, found in gastric MALT lymphomas, have never been reported [1, 17]

The association with a Lyme borreliosis has been reported in Asian cases, hypereosinophilia has been frequently described [7]. Autoimmune diseases are rarely associated contrary to systemic forms, [4].

The prognosis is good, it is known to be an indolent form, with a Survival at 5 years> 95% (4-6). but recurrences are frequent, around 60%, with a median delay of 16 months [18]. Extra-cutaneous spread, and mortalities linked to the disease are even rarer (8.5%). Extracutaneous extensions are typically preceded by transformation into large cell lymphoma [4, 19, 20].

The therapeutic choices are wide: surgery, radiotherapy and topical, intralesional or systemic treatment, rituximab as monotherapy for more extensive lesions, chemotherapy with multiple agents is rarely appropriate. Partial to complete remission has been reported with the use of rituximab as an infusion or intra-lesion [21, 22]. The choice of treatment takes into consideration, the indolent nature of the condition, the patient's age and background, the extent of the lesions, and recurrences.

Conclusion

Primary cutaneous marginal zone B-cell lymphoma is an indolent B-cell lymphoma, typically characterized by frequent skin recurrences and exceptional systemic extension.

The real challenge is the diagnosis, given the clinical polymorphism and the deceptive forms, as well as the great similarities and histological nuances

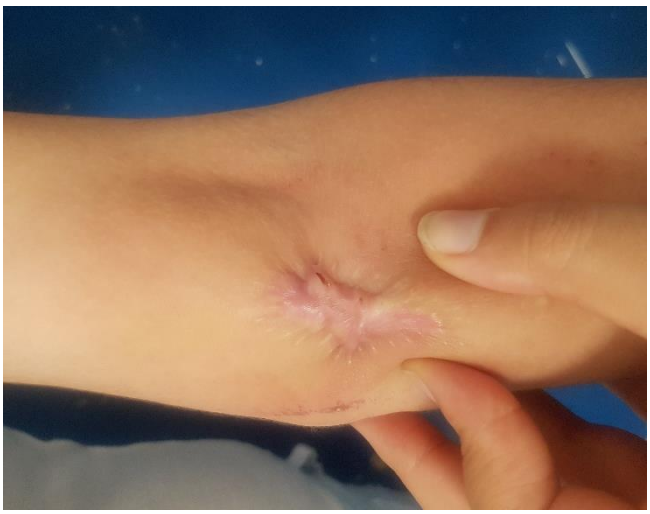

Fig 1: subcutaneous tumor at the level of the left elbow, poorly limited, without infiltration of the adjacent skin, where, in addition to the scars of old surgical procedures, an eczematiforme reaction is observed.

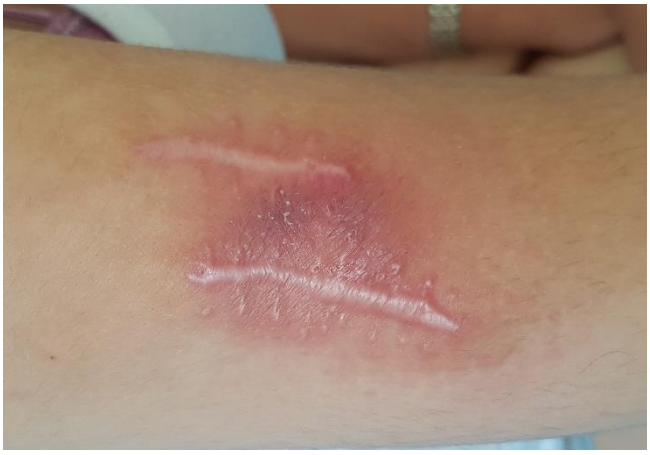

Fig 2: at the level of the arm, the lesion is not visible, two hypertrophic scars were objectified, associated with an eczematiforme erythematous plaque.

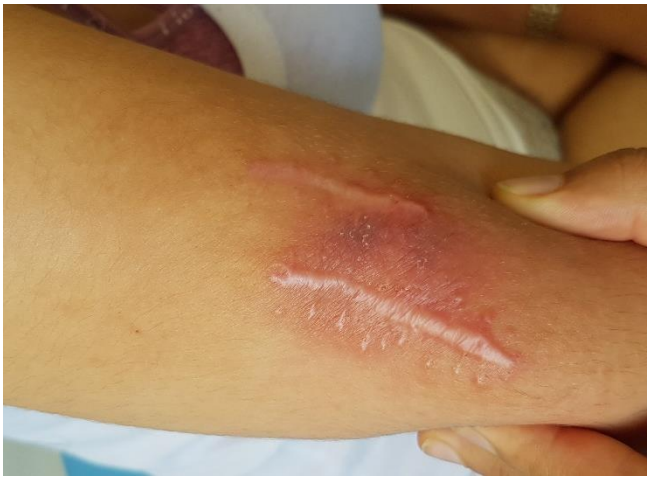

Fig 3: We can palpate the subcutaneous tumor, which is ill-defined and painless

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