

PRIMARY CUTANEOUS LYMPHOMA.

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Approximately 25 percent of all NHL cases will present at an extranodal site without systemic involvement. The skin is the second most common primary extranodal site, second in frequency only to the gastrointestinal tract. The overall incidence of primary cutaneous lymphomas in Western countries is estimated to be 0.5 to 1 case per 100,000 people annually. The clinical and histologic diagnosis of cutaneous lymphoproliferative disorders is one of the most vexing issues in dermatology and dermatopathology, despite significant advances in their classification, pathogenesis, and treatment. The average delay between initial presentation and the ultimate diagnosis of mycosis fungoides, the most common primary cutaneous T cell lymphoma, is six years. During this period, patients typically undergo numerous skin biopsies, which, in the absence of definite histopathologic criteria for early mycosis fungoides, may be interpreted by the pathologist as “atypical lymphocytic infiltrate” or “atypical lymphocytic proliferation.” This nonspecific but potentially serious diagnosis is often a source of anxiety and frustration for both the patient and the clinician. — The term “atypical lymphocytic infiltrate” describes the histologic finding of a dermal infiltrate of atypical lymphocytes admixed with cytologically banal, reactive-appearing lymphocytes in a pattern that is suggestive of lymphoma or leukemia and is generally used when the pathologist cannot reliably differentiate a reactive from a malignant lymphoproliferative disorder on histopathologic. However, despite histologic clues, immunohistochemical staining patterns, and molecular data, pathologists are often left without a definitive diagnosis. In such situations, the best approach is to describe the salient histopathologic features and provide the clinician with an extensive differential diagnosis based upon the synthesis of the various findings. In addition, a direct discussion of the case with the clinician may be extremely helpful to both parties, especially if the possibility of a lymphoproliferative disorder was not clinically suspected.

Following these steps will eventually lead sooner or later to diagnosis and classification into B or T cells lymphoproliferations.

Regarding B cells lymphoproliferative disorders with primary cutaneous involve, we consider three aspects (primary cutaneous follicle center lymphoma, primary cutaneous large B cell lymphoma, leg type and primary cutaneous marginal zone lymphoma in which initial staging is mandatory the absence of any other systemic involve/lesion.

These clinical entities suggest similarities with various steps in B cells ontogenesis, similarities sustained by clinical, histological, immunophenotype and molecular aspects for each lesion. The prognosis, evolution and therapeutic approach varies.

Skin-based lymphoma of T cell origin includes besides mycosis fungoides, and Sezary syndrome, also CD 30+ lymphoproliferative disorders, (L/LTA), extranodal NK/T cell lymphoma, nasal type, subcutaneous panniculitis-like T cell lymphoma and primary cutaneous aggressive, epidermotropic CD8+ T cell lymphoma, cutaneous $\gamma\delta$ T cell lymphoma, primary cutaneous CD4+ small/medium size lymphoma.

In conclusion primary cutaneous lymphoproliferative disorders are not so rare, are difficult to diagnose and includes aggressive form which needs rapid diagnosis and treatment, among with indolent forms that take years of evolution until diagnosis.