DERMATOLOGY PRACTICAL & CONCEPTUAL
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Review: Skin Lymphoma. The Illustrated Guide, 4th edition, by Lorenzo Cerroni

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Citation: Milette F. Review: Skin Lymphoma. The Illustrated Guide, 4th edition, by Lorenzo Cerroni. Dermatol Pract Concept. 2014;4(4):17. http://dx.doi.org/10.5826/dpc.0404a17

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Review by François Milette

Lorenzo Cerroni is the sole author of the fourth edition of this wonderful book on a most difficult, complex and confusing subject: cutaneous lymphoma. The former three editions were co-authored by Helmut Kerl, who recently retired, and Kevin Gatter, who now focuses on his work on bone marrow diagnosis. I do not doubt for a single second that Dr. Cerroni's task has been, as stated by him, "Herculean."

The book is remarkably organized and presented. Its reading is easy and its iconography outstanding, both clinically and histopathologically. It is very "user friendly," its size being reasonable (425 pages). Most of all (and this is the main reason for buying a book!) it is easily, efficiently and usefully consulted when one is facing a difficult case. For many years I have used regularly the former editions of this guide and I am certain I will continue to use this new edition for many more years. Last and not least, many chapters are supplemented with very instructive teaching cases that illustrate the never overestimated importance of clinico-pathological correlations and of open-mindedness in dealing with cutaneous lymphomas.

The structure of Cerroni's book is similar to that of the preceding editions. The various lesions are grouped under the following headings: (1) NK/T-cell lymphomas, (2) B-cell lymphomas, (3) Lymphomas in immunosuppressed individuals, (4) Leukemias and precursor hematologic neoplasms, (5) Hodgkin's lymphoma, (6) Lymphomas in children and adolescents, (7) Pseudolymphomas and (8) "atypical lymphoid proliferation." In this last section, neoplasms and lesions hard to categorize are discussed. These neoplasms either do not fit into a defined category or their benignancy/malignancy is undecidable. Although this classification is not entirely coherent, being based sometimes on histological, sometimes on clinical, sometimes on prognostic data, it certainly has some pertinence in relation with day-to-day practice.

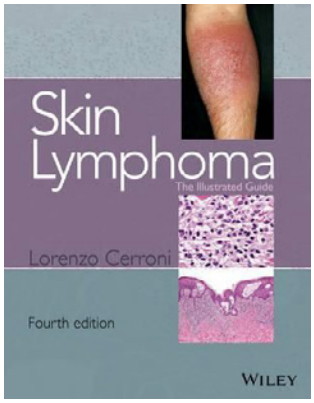

Figure 1. Cerroni L. *Skin Lymphoma. The Illustrated Guide*. 4th ed. Wiley-Blackwell 2014. ISBN 978-1-118-49249-9. 425 pages. $219.95

The subtle differences between the table of content of this new edition and that of the third edition illustrate well the subjectivity of any classification. For example, intravascular large cell lymphomas of the B, T and CD30 lineages are included in section 2 on B-cell lymphomas, whereas in the third edition they were classified, together with lymphomas in immunosuppressed individuals, in section 3 devoted to "Other cutaneous lymphomas" (a section that no longer exists, having been replaced by a new section 3 devoted to

lymphomas [B-cell and T-cell] developed in immunosuppressed individuals). For these types of lymphomas, the intravascular ones, vascular dissemination of cells was considered determinant of their conceptualization. Perhaps, in a later edition, these intravascular variants will be redeployed in the sections corresponding to their cell types.

As coauthor of a book on a single type of cutaneous lymphoma [1] and detractor of imprecision in language, I know well how difficult it is to bring order of any kind in the confused and controversial field of cutaneous lymphomas. These conceptual problems are admitted by Dr. Cerroni, as he recognized the existence of "a grey zone between clearly benign and clearly malignant neoplasms, a cloudy area where conventional definitions and criteria do not always work." Everybody would admit the existence of these cloudy areas and grey zone, but it is no reason to accept definitions that are non-definitions, definitions that defy understanding, such as the following definition of "indolent CD8+ lymphoid proliferation of the ear (face)" (page 166): *a CD8+/ CD4- variant of CD4+ small-medium T-cell lymphoma that is not confined to the ears or face*! There is a name for such a statement: oxymoron.

And what should one think of this "definition" of mycosis fungoides to be found on page 11:

> Mycosis fungoides is the most common type of cutaneous lymphoma, representing almost 50% of all lymphomas arising primarily in the skin. It is defined as [emphasis mine] a tumor composed of small/medium-sized, epidermotropic T-helper lymphocytes (but T-cytotoxic variants are not uncommon and tumor cells may be medium/large in advanced stages).

After comparing this definition with that proposed in our opus [1], the reader is invited to conclude:

> Mycosis fungoides is a disease systemic from the outset, neoplastic, malignant, composed of T-lymphocytes (i.e.: a lymphoma) with affinity special to the skin, with presentation protean, clinically, histopathologically, and biologically, of cause unknown.

Although our definition can be and has been criticized as overly inclusive, the protean character of mycosis fungoides is acknowledge by Dr. Cerroni, who lists the following variants of MF, clinical and histological: parapsoriasis, folliculotropic (with or without follicular mucinosis), syringotropic, localized pagetoid, unilesional, granulomatous, slack skin, erythrodermic, interstitial, poikilodermic, hypopigmented, hyperpigmented, purpuric, papular, bullous, anetodermic, PLEVA-like, "invisible," etc.

Incidently, one should rejoice since it is clearly admitted at last (pages 34-35) that parapsoriasis, be it of the small or large plaque type, is mycosis fungoides even though the author continues to insist on the fact that "regardless of the academic discussion, it is important to underline that patients with small plaque parapsoriasis should not be aggressively treated." This is evident and true not only for small-plaque parapsoriasis but also for any clinically indolent lymphoma, even for any indolent neoplasm of any type.

Where is the limit to the protean character of MF to be set? Considering unilesional T-cell lymphoma, for instance, is it justified, as does Cerroni, to subdivide it into (1) pagetoid reticulosis (with massive epidermotropism), (2) CD4+ small and medium T-cell lymphoma (with very little or no epidermotropism and diffuse dermal infiltration), (3) "genuine" unilesional MF (defined clinically), and (4) solitary MF with large cell transformation (Teaching case 2.2, page 63), a supposedly "worrying feature" even if the patient remains in complete remission, one year after treatment? And is it reasonable to exclude from the spectrum of MF the leukemic variant of it (Sézary syndrome) on the basis of the presence of more than 1,000 malignant circulating cells per cubic millimeter of blood and of purportedly specific molecular characteristics? Isn't it very probable that such molecular or genetic changes exist for each and every variant accepted as parts of the spectrum of MF? If one insists on genetic profiling of tumors, isn't it to be feared that there will soon be as many lymphomas as there are patients harboring lymphomas? These questions merit reflection.

Many other concepts addressed in this otherwise wonderful book could be discussed at length, but that would clearly take us beyond the limits of a book review. As a conclusion, I would say that although this book continues to be plagued with some of the conceptual weaknesses surrounding the study of cutaneous lymphoma, it is a very good book and its author cannot be held responsible for all the "tossed salads of words" that for years have been feeding the minds of hematopathologists in search of, not proper diagnosis, but of precise prognostication.

In a word, I warmly recommend this book that deals exhaustively with a very difficult subject. It can be very useful on a day-to-day basis but certainly has to be read with discernment.

Reference

1. Ackerman AB, Denianke K, Sceppa J, Asgari M, Milette F, Sanchez J. *Mycosis Fungoides: Perspective Historical Allied with Critique Methodical for the Purpose of Illumination Maximal. Atlas and Text*. New York: Ardor Scribend, Ltd., 2008.