Review

A review of the solitary cutaneous T-cell lymphomas

Cutaneous T-cell lymphomas (CTCL) account for almost 65-92% of all cutaneous lymphomas, many of which usually present with multiple lesions. However, a number of well-recognized and rare types of CTCL, including mycosis fungoides, can present in isolated fashion. These solitary lesions often run a relatively indolent clinical course but often pose diagnostic difficulties. We review histopathologically challenging solitary cutaneous T-cell lymphomas, including criteria for diagnosis, clinical course and prognosis, particularly for primary cutaneous CD4+ small/medium pleomorphic lymphoma and indolent CD8+ lymphoid proliferation of acral sites. In addition, we suggest an algorithm and nomenclature to aid in the diagnosis of such problematic lesions.

Keywords: solitary mycosis fungoides, pagetoid reticulosis, indolent CD8+ lymphoid proliferation of acral sites, primary cutaneous CD4+ small/medium pleomorphic lymphoma

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Primary cutaneous T-cell lymphomas present in the skin with no evidence of extra-cutaneous involvement.1 Cutaneous T-cell lymphomas (CTCL) account for almost 65–92% of all cutaneous lymphomas,2 of which, mycosis fungoides (MF) is the most common and characteristically presents with multiple lesions. However, solitary MF, including and distinct from pagetoid reticulosis (Woringer-Kolopp), is well documented;3 similarly, other well-characterized forms of lymphoma can present as an isolated lesion. Finally, a number of increasingly recognized rarer CTCL commonly present with an isolated lesion, including CD4+ small/medium pleomorphic T-cell lymphoma (SMPTCL) and indolent CD8+ lymphoid proliferation of acral sites.4 Although it seems that these isolated lesions run a relatively indolent clinical course, they continue to pose diagnostic difficulty.

In this review, we present an overview of the solitary cutaneous T-cell lymphomas that, in our opinion, present the greatest challenge in diagnosis and that have been emerging over the last 5 years. We have excluded some of the more clinically aggressive tumors that may present as solitary lesions as they tend to be well recognized and histopathologically and immunophenotypically distinct. We discuss the criteria for diagnosis, particularly for primary cutaneous CD4+ small/medium pleomorphic lymphoma and indolent CD8+ lymphoid proliferation of acral sites. Finally, we suggest an algorithm to aid in the diagnosis of such problematic lesions.
Review

Cutaneous lymphoma classification and review

The recent détente and concord between the World Health Organization (WHO) and European Organization for the Research and Treatment of Cancer (EORTC) classifications acknowledges the unique features of primary cutaneous lymphomas, which differ greatly from the nodal and other extranodal lymphomas. An accurate diagnosis of primary cutaneous lymphoma requires the correlation of the clinical findings with the microscopic, immunophenotypic and genetic features. Given that the majority of lymphomas usually present with multiple lesions it is difficult to make a diagnosis of a specific cutaneous lymphoma when faced with the clinical presentation of a solitary lesion. Moreover, in the absence of multiple lesions the surgical pathologist is frequently in difficulty in determining whether an infiltrate is neoplastic or reactive.

To date, of the solitary cutaneous T-cell lymphomas reported in the literature, approximately 166 cases represent MF, 231 represent SMPTCL, 22 represent indolent CD8+ lymphoid proliferation, and 62 represent pagetoid reticulosis (Table 1).

Solitary MF

MF accounts for almost 50% of all primary cutaneous lymphomas and is characterized by multiple patches and plaques. Indeed, given that the finding of multiple patches and plaques is a defining feature it is somewhat contradictory to consider a solitary variant. Nevertheless, since 1981, 166 solitary cases of MF have been described that are distinct from pagetoid reticulosis, including 10 of the folliculotropic variant (follicular MF) and 9 cases of syringotropic MF.

Clinical features

Solitary MF, clinically and histopathologically resembles classic MF. Clinically, it presents as a single erythematous scaly patch or plaque, in non-sun-exposed areas. Solitary MF has been reported to have an excellent prognosis, although recurrence of treated lesions has been reported six of which occurred distant from the site of the original lesion (Table 1). Two of these distant recurrences presented as multiple lesions, but in both cases, there was complete resolution with further treatment and no further disease at follow-up. The other four cases presented as a unilesional recurrence. In addition, four cases of solitary MF have progressed to further cutaneous involvement, three of which had large cell transformation.

Solitary lesions of follicular and syringotropic MF are also clinically and pathologically indistinguishable from lesions seen in classical disease. Solitary folliculotropic MF presents as an infiltrated patch, plaque or an isolated area of follicular papules without prominent scaling, usually in the head and neck area. Solitary syringotropic MF usually presents with a single erythematous, indurated plaque, often studded with small pinhead-sized papules, in the trunk, hip and thigh area, although cases have been reported in the head and neck area. Alopecia is a prominent feature in both cases. There is still uncertainty about the relationship between these two MF subtypes, but they often co-exist. When compared with classic MF, folliculotropic MF is typically associated with a poorer prognosis. This is sometimes, but not always the case for solitary lesions. Syringotropic MF, however, has a prognosis similar to that of classic MF. Out of the nine solitary cases of syringotropic MF reported, one has shown recurrence of the lesion distant from the original site.

Finally, the report of an EORTC workshop concerning granulomatous MF and granulomatous slack skin (GSS) detailed one solitary example of GSS. Recently, at the senior author’s institution, a case of GSS presented with a single pendulous axillary skin fold, which was followed.

<table>
<thead>
<tr>
<th>Cutaneous lymphoma type</th>
<th>Total cases</th>
<th>Cases with cutaneous/systemic progression</th>
<th>Cases with lesion recurrence post-treatment*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Solitary MF</td>
<td>166</td>
<td>4 (Cutaneous spread, 3 × large cell transformation)</td>
<td>15 (six distant from original site)</td>
</tr>
<tr>
<td>Pagetoid reticulosis (Woringer-Kolopp)</td>
<td>62</td>
<td>1 (Cutaneous spread)</td>
<td>8 (1 distant from original site)</td>
</tr>
<tr>
<td>SMPTCL</td>
<td>231</td>
<td>5 (Extracutaneous, one died of lymphoma)</td>
<td>12</td>
</tr>
<tr>
<td>Indolent CD8+ lymphoid proliferation of acral sites</td>
<td>22</td>
<td>0</td>
<td>4 (one distant from original site)</td>
</tr>
</tbody>
</table>

*Recurrence occurred at the same site of the original lesions unless stated otherwise.
A recent report by Ally et al. of 15 cases suggested that the observation of solitary MF might be expected, albeit exceptional, in which the disease is simply found at an uncommonly early stage. The unsurprising excellent prognosis of such cases indicates that therapy should aim to be curative.

**Histopathologic features**

In solitary MF, microscopic sections show a superficial lymphoid infiltrate of epidermotropic and atypical lymphocytes. Atypia is usually more pronounced in the epidermotropic lymphocytes than those in the dermis. Other histopathologic features include lining up of atypical lymphocytes at the dermoepidermal junction, Pautrier microabscesses and fibrotic changes in the papillary dermis, none of which is specific for the diagnosis of MF (Fig. 2). Indeed, Cerroni et al.’s series of 20 cases lacked many of these features. Folliculotropic and syringotropic MF are characterized by infiltration of atypical lymphocytes within and around the hair follicle and/or eccrine epithelium, sometimes with follicular mucinosis. However, atypia can be minimal and distinction from idiopathic prior to follicular mucinosis is problematic; follow-up with repeated biopsies may be required. Thus, in short, the histopathologic features of solitary lesions of MF mimic the features of the classic disease.

In the context of a solitary lesion, perhaps the burden of proof for the diagnosis should be particularly stringent; in Ally et al.’s series of 15 patients there were at least two of the following: morphological and cytological features of MF, loss of T-cell associated antigens, T-cell clonality and appropriate clinical appearance.

**Immunocytochemistry**

Immunophenotypically, solitary MF, like classic MF, is usually a neoplasm of CD4+ T-helper cells, although CD8+ and double negative CD4−/CD8− variants have been described. In addition, there may be variable antigen loss of CD2, CD5 and CD7. These immunophenotypes do not seem to affect prognosis.

**T-cell receptor gene analysis**

T-cell receptor (TCR) gene rearrangements are detected in approximately 57–71% of cases of early MF, so solitary lesions might be expected to have a similar or perhaps lower clonal prevalence. Out of the 166 cases of solitary MF published, approximately 76 were tested for TCR clonality, of which 45 cases had a clonal TCR rearrangement. The detection of monoclonal TCR rearrangements does not appear to be of prognostic impact.

**Pagetoid reticulosis (Woringer-Kolopp)**

In 1939, Woringer and Kolopp reported a solitary plaque on the forearm of a 13-year-old boy, which resembled MF. Since then, the term pagetoid reticulosis was introduced, to recognize the similarity between the atypical epidermotropic cells in this disease and the intraepidermal adenocarcinomatous cells of Paget’s disease of the nipple. In the 2005, WHO/EORTC classification of cutaneous lymphomas, pagetoid reticulosis is classified as a...
variant of MF, despite having distinct clinical and histopathologic features.14,34–36

Clinical features

The typical lesion of pagetoid reticulosis is a solitary, slow growing, well circumscribed, psoriasisform or hyperkeratotic plaque1,20,37 (Fig. 3). In some cases, patients present with multiple lesions.38 Pagetoid reticulosis typically affects the distal extremities,39 although, lesions on the trunk39 and even the tongue36 have been described. Pagetoid reticulosis can affect any age group including young children40 and usually has an indolent clinical course, although there is potential for dissemination and malignant behavior.20,41 Some cases have been associated with the development of MF several years after the development of pagetoid reticulosis,1,38,42 highlighting the need for long-term follow-up. In contrast to solitary MF, extracutaneous dissemination or disease-related death has never been reported.1

Histopathologic features

Pagetoid reticulosis is characterized by epidermal hyperplasia with parakeratosis, prominent acanthosis and florid epidermotropism of atypical lymphocytes typically scattered throughout the epidermis1,20 (Fig. 4A). It is this pattern of epidermotropism that is characteristic, and despite the high number of intraepidermal lymphocytes, Pautrier microabscesses are absent or rare in the authors’ experience. The dermal infiltrate in pagetoid reticulosis consists of reactive lymphocytes and histiocytes. In contrast, MF is characterized by atypical lymphocytes above and below the dermal-epidermal junction.38 In observing a pagetoid reticulosis pattern, attention to clinical features is required to distinguish these cases from lymphomatoid papulosis (LyP) type D (Fig. 4B) or the more recently described variant of LyP associated with 6p25.3 rearrangements, rare cases of MF with this pattern and aggressive epidermotropic CD8+ T-cell lymphoma.43 In addition, microscopic distinction of pagetoid reticulosis from some cases of CD8+ pityriasis lichenoides et varioliformis acuta cases may be challenging, especially when only sparse clinical data are provided.44

Immunocytochemistry

In a recent review by Mourtzinos et al.,45 the neoplastic T-cells in pagetoid reticulosis were CD8+/CD4− in 53% (Fig. 4C), CD4+/CD8− in 36% and CD4−/CD8− in 11% of cases. Furthermore, approximately 47% of pagetoid reticulosis cases showed strong and extensive CD30 expression; in such cases the distinction from lymphomatoid papulosis is particularly germane. High CD30 expression is uncommon in patch stage MF, and is largely found in later stages and/or large cell transformation.46 To our knowledge, it has never been reported in cases of solitary MF. The immunophenotype in pagetoid reticulosis appears to be prognostically irrelevant.45

T-cell receptor gene analysis

Previous studies have stated that the majority of cases of pagetoid reticulosis reveal clonal rearrangements on analysis of TCR genes.20,38,41 However, approximately 13 cases in which pagetoid reticulosis presented with a solitary lesion were tested for clonality, and of those only 6 were identified as clonal.31,38,47–49

Primary cutaneous CD4+ small/medium pleomorphic T-cell lymphoma (SMPTCL)

SMPTCL has been credited to account for approximately 3% of all primary cutaneous lymphomas and is listed as a provisional entity in the WHO-EORTC classification of cutaneous lymphomas.1 With recent increased recognition, however, in the authors’ experience it is probably the most common CTCL. To date, many series of cases have been reported50–55 the largest of which included 133 patients with solitary lesions.51 The relatively recently recognized entity of CD8+ lymphoid proliferation of acral sites56 has been speculated to be a phenotypic variant of SMPTCL.57,58 It is a particularly
Pagetoid reticulosis is characterized by epidermal hyperplasia with parakeratosis, prominent acanthosis and florid epidermotropism of atypical lymphocytes typically scattered throughout the epidermis (A, hematoxylin/eosin, ×100), similar to that seen in lymphomatoid papulosis type D (B, hematoxylin/eosin, ×100). The neoplastic T-cells in pagetoid reticulosis are often CD8+/CD4(−) (C, CD8 immunostain, ×100).

contentious neoplasm, both nosologically and diagnostically.

Clinical features

SMPTCL usually presents with a solitary plaque or tumor, on the face, neck or upper trunk\textsuperscript{1,51} (Fig. 5). In total, 231 solitary lesions of SMPTCL have been described. Less commonly, it may present with multiple lesions\textsuperscript{50,52,53} SMPTCL usually occurs in patients aged between 50 and 60 years old, although paediatric cases have also been described\textsuperscript{50,51,53} Cases presenting as solitary skin lesions typically have an excellent prognosis\textsuperscript{50,51,53}, although some cases have experienced recurrence of treated lesions\textsuperscript{50,51,55–55} or even extracutaneous spread and death\textsuperscript{52} (Table 1). The latter study describes five cases in which there followed an aggressive clinical course, all of whom had nodules >5 cm in size. One of these cases co-expressed CD20, there were technical problems with CD4 in another and at least focal loss of CD4 was observed in three, one of which transformed to large cell lymphoma. However, no immunophenotyping for follicular helper T-cell expression was performed and perhaps the diagnosis in these cases may be questioned. The WHO-EORTC classification cites a 5-year survival of only 70% although subsequent reports – and most authorities – now accept the prognosis is much better\textsuperscript{1} Baum et al.\textsuperscript{50} described waxing and waning of lesions. The pathologic criteria for diagnosis have evolved erratically rather than been systematically defined, and framed within
Fig. 6. SMPTCL presents with a dense dermal infiltrate consisting predominantly of small/medium pleomorphic T-cells, often with hyperchromatic angulated nuclei, which exhibit a tendency to extend to the subcutis (A&B, hematoxylin/eosin, ×40 and ×100, respectively). Rosettes of neoplastic T-cells are highlighted by PD-1 (C, PD-1, ×200) and a heavy B-cell population is oftentimes present (D, CD20, ×40).


Histopathologic features
SMPTCL almost always presents a dense infiltrate of predominantly small/medium pleomorphic T-cells packing the dermis, with a tendency to extend to the subcutis. The cells have hyperchromatic angulated nuclei (Figs. 6A,B). A small proportion of large cells may be observed (by definition, <30%).1 Epidermotropism may be present focally54 and in some cases there are a considerable number of small reactive lymphocytes.

Immunocytochemistry
By definition, the phenotype in SMPTCL is CD3+, CD4+, CD8−, CD30−.1 However, CD8+ variants have been described, some of which are lacking detailed analysis.51 One CD8+ variant was described to occur on the foot and appeared to have an indolent clinical course.54 It is now recognized that SMPTCL is a neoplasm of follicular T-helper cells and, accordingly, expresses PD-1, ICOS, CXCL-13, bcl-6 and CD10 (Fig. 6C).

Interestingly, these markers are not consistently expressed to the same degree – perhaps reflecting differing specificities, but also that immunophenotype at extranodal sites does not always mirror the nodal findings.59 A few authors stress the finding of rosettes of neoplastic T-cells highlighted by the immunophenotype (Fig. 6C) and often surrounding a B-cell infiltrate (Fig. 6D).55 The follicular T-helper cell phenotype is believed to underlie the prominent B-cell population that is a consistent feature of this tumor.53,55

Problems of definition and diagnosis
The usual difficulty when faced with this tumor is deciding whether the dense but quite polymorphous infiltrate is reactive or neoplastic. The concept of ‘nodular T-cell pseudolymphoma’ is a source of further obfuscation. In the authors’ view, terms such as pseudolymphoma (PSL) or nodular T-cell pseudolymphoma are unhelpful and misleading.60 Many literature reports attest to the varied conditions that can mimic a lymphomatous infiltrate, including drug ingestion, Jessner’s lymphocytic infiltrate, arthropod reactions, lymphomatoid keratosis,
herpes infection, contact dermatitis, tattoo and pseudolymphomatous folliculitis (PLF).60–64
Therefore, T-cell pseudolymphoma sui generis does not exist; instead, the plethora of conditions reported should simply warn the wary pathologist of mistaking a diagnosis for lymphoma in certain settings. In addition, no consensus definition of ‘nodular T-cell pseudolymphoma’ has been reported to the authors’ knowledge. Most publications of this ‘entity’ date before the year 2000, and have not been subject to modern techniques for assessing clonality or follicular T-helper cell phenotyping.60 One recent report did seek to compare PSL with SMPTCL but included a PSL in the latter as they were deemed to be ‘identical’, prejudging the very comparison.65 In fact, any attempted comparison between SMPTCL and ‘nodular T-cell pseudolymphoma’ is problematic as they are likely one and the same. If no causative stimulus is identified for an infiltrate that has at least some features suggestive of lymphoma to the pathologist then, in our opinion, the diagnosis should be ‘lymphoid infiltrate of uncertain nature’ rather than ‘T-cell pseudolymphoma’. Since the demonstration of a follicular helper T-cell phenotype, it is reasonable to insist this is a required diagnostic criterion for SMPTCL, although which are the more reliable markers of this phenotype is not entirely clear. Our group examined whether follicular helper T-cells were present within the infiltrates of a broad range of reactive dermatoses, including drug reactions. In these cases, the expression of PD-1, ICOS and CXCL-13 was always low, and lacked rosettes.66 Clonality is observed in around 60–70% of cases but Grogg et al.55 assert that a dominant T-cell clone should be an essential criterion. Although, clonal rearrangements of the TCR are highly suggestive of lymphoma, it is accepted that reactive conditions may also exhibit T-cell monoclonality,64 making it unsatisfactory to use this as a definitive tool. In the authors’ view, the typical microscopic features should be combined with widespread expression of PD-1 exhibiting a rosette pattern, and at least one more of either bcl-6, ICOS, CXCL-13 or CD10; loss of a T-cell antigen and/or T-cell clonality are additional supportive features if present. We suggest that in such cases, arising in the clinical context of features typically as depicted in Fig. 5, it is more than reasonable to assert a diagnosis of T-cell lymphoma, albeit one which is almost certainly low-grade.

The heavy B-cell population oftentimes present (Fig. 6D) underlies a further common diagnostic problem viz. a solitary lesion having a mixed T- and B-cell infiltrate, with relatively low-grade atypia, causes difficulties for pathologists in determining whether it is a T or B-cell malignancy. Marginal zone lymphoma (MZL) and T-cell rich B-cell lymphoma (TCRB) – each often with heavy T-cell populations – enter the differential diagnosis. Both are outside the scope of this review but, with respect to MZL, attention should be paid to plasmacytoid differentiation and the presence of monoclonal population with in-situ hybridization to light chains. TCRB is not afforded a separate primary cutaneous category in the WHO-EORTC classification, and while convincing reports exist67 it is interesting to postulate whether at least some reported examples reflect confusion between variants of MZL and SMPTCL.

Finally, since the observation of the follicular T-helper phenotype, several reports have emerged of a putative entity, ‘follicular T-helper cell lymphoma’ (FTHCL), and this has included solitary and multiple forms.68 The relationships between SMPTCL, FTHCL and angioimmunoblastic lymphoma (AIL) – also a helper T-cell neoplasm – remain to be elucidated but thus far it appears that solitary tumors, as usually seen with SMPTCL, have an excellent
Fig. 8. Indolent CD8-positive lymphoid proliferation exhibits a dense, diffuse, dermal lymphoid infiltrate with a Grenz zone (A&B, hematoxylin/eosin, ×100). Rare cases with one or two Pautrier collections have been reported (C, hematoxylin/eosin, ×200) and the infiltrate is characteristically monotonous (D, hematoxylin/eosin, ×100).

prognosis, while in the presence of multiple lesions, as seen in FTHCL and AIL, predictions should be more guarded. Such preliminary findings should obviously be taken into account while planning appropriate therapy.

T-cell receptor gene analysis

In one of the largest published series on SMPTCL, monoclonal rearrangements of the TCR were demonstrated in 60% of cases.51 Some studies reported even higher proportions.53

Indolent CD8+ lymphoid proliferation of acral sites

To date, only 22 solitary lesions of indolent CD8+ lymphoid proliferation have been described. A recent report described six patients with a wider constellation of clinical sites and microscopic features than previously recognized.69 Given the occurrence on the foot and hands, it was suggested that indolent CD8+ lymphoid proliferation of acral sites was a more apposite name, although the ear remains the most common location (Fig. 7A).

Clinical features

Clinically, they present as erythematous papules or nodules, often on the ears, but two have occurred on the nose,70,71 and also hands and feet (Figs. 7B,C). It appears to have a predilection for males, and has a mean incidence age of 54 years.58 Lesion recurrence post-treatment has been reported, but there is no evidence as yet to suggest that this entity has an aggressive nature.4,57,58,69 Nevertheless, at least one patient continues to develop small papules at various acral sites.69

Histopathologic features

Indolent CD8+ lymphoid proliferation presents with a dense, diffuse, dermal lymphoid infiltrate, separated from the epidermis by a zone of sparing (Figs 8A,B). Foci of epidermotropism and even an occasional Pautrier microabscess have been reported (Fig. 8C), but follicular and sweat apparatus involvement are never seen.69 The infiltrate consists of atypical medium-sized lymphocytes with a small minority population of reactive lymphocytes (mainly B-cells). The
lymphocytes are described as pleomorphic in some cases, but monomorphic in most other reports. The infiltrate is characteristically monotonous in the authors’ experience (Fig. 8D) and differs from the more obviously polytypic infiltrate of SMPTCL. All cases of indolent CD8+ lymphoid proliferation have a CD8+ phenotype, by definition. Beltraminelli et al. has suggested that, based on morphology, indolent CD8+ lymphoid proliferation best fits into the WHO/EORTC category of SMPTCL and that it should be considered a phenotypic variant of SMPTCL. Swick et al. and Kempf et al. agreed with this suggestion and Geraud et al. described another case of this entity and termed it ‘primary cutaneous CD8+ small/medium sized pleomorphic T-cell lymphoma, ear type’. However, a recent series by Greenblatt et al. found a complete absence of expression of follicular helper T-cell markers in four cases, and the tumor cell morphology – in most publications – seems quite distinct from SMPTCL.

Locally directed therapy is the treatment of choice. Wider recognition of this entity is required to prevent unnecessarily aggressive chemotherapy for what is otherwise often labelled Peripheral T-cell Lymphoma NOS, a category usually interpreted as signifying life-threatening disease.

Immunocytochemistry

By definition tumor cells express CD8, diffusely and strongly in all cases so far, and might have loss of expression of one or more T-cell associated antigens. In most examples, there is a low proliferative fraction, and, although the neoplastic cells express TIA-1, they are usually granzyme B and perforin negative. A few exceptions have been recently documented with higher than usual (30–40%) Ki-67 index and tumors positive for granzyme B.

T-cell receptor gene analysis

Of the 22 described cases of indolent CD8+ lymphoid proliferation, 14 were analyzed for TCR rearrangements and 13 were found to be clonal. Other lymphomas reported occurring as solitary lesions

A wide variety of lymphomas have been reported as presenting with a solitary lesion. These include anaplastic CD30+ large cell lymphoma, which commonly presents as a single tumor, but clearly

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Fig. 9. Diagnostic algorithm for solitary cutaneous T-cell lymphomas. MF, mycosis fungoides; PR, pagetoid reticulosis; ICC, immunocytochemistry; SMPTCL, primary cutaneous CD4+ small/medium pleomorphic T-cell lymphoma; *Further investigations required to clarify disease or etiology; #of atypical lymphocytes.
spread from a systemic primary needs to be excluded; lymphomatoid papulosis, although such cases reflect the initial presentation, as a single lesion is inevitably followed by crops of papules; subcutaneous panniculitis-like T-cell lymphoma, and CD20+ cutaneous T-cell lymphoma NOS. A repeated observation in the majority of such cases is the excellent prognosis.

**Recommendations**

On the basis of the literature review presented herein, we have proposed a system for the diagnosis of some of lymphomas that might present as a solitary lesion, which involves clinical, microscopic, immunocytochemical and molecular biologic features (Fig. 9). The proposed algorithm summarizes many of the points discussed.

**Conclusion**

A variety of cutaneous T-cell lymphomas can present as a solitary lesion, and regardless of type these almost always have a better clinical outcome than the more common multiple counterpart; treatment can aim, therefore, to be curative. Nevertheless, follow-up is warranted. We propose a system to aid in the classification of these lesions based on a combination of clinical, histopathologic, immunocytochemical and molecular biologic features.

**References**


