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Review

Primary Cutaneous B-Cell Lymphomas

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Abstract

Primary cutaneous B-cell lymphomas (PCBCL) are a heterogeneous group of mature B-cells neoplasms that present in the skin without evidence of nodal or systemic involvement. The clinical and pathologic features of PCBCL differ significantly from the equivalent nodal lymphomas. Three main subtypes of PCBCL are recognized by the 2016 revised WHO classification. Studies have shown that PCBCLs are characterized by distinct immunophenotypic features, chromosomal aberrations and gene rearrangements which provide further support for their classification as separate entities from their nodal types.

Keywords
Skin diseases, lymphoma, B-lymphocytes, diffuse large B-cell lymphoma, marginal zone lymphoma, follicle center cell lymphoma

Identity

Lymphoma is classified as primary cutaneous lymphoma (PCL) when the malignant lymphocyte proliferation is limited to the skin with no involvement of lymph nodes, bone marrow or viscera at diagnosis. PCLs are the most frequent extra-nodal lymphomas, with incidence around 10 cases per million individuals per year, from which 25-30% are primary cutaneous B-cell lymphomas (PCBCL) (Kempf W et al 2014 Mar). PCBCLs are characterized by great biological and clinical variability among its various subtypes (Sokotowska-Wojdyto 2015). The classification was revolutionized by the consensus statement between the World Health Organization (WHO) and European Organization for the Research and Treatment of Cancer in 2005. The distinct cutaneous B-cell lymphoma entities are discussed here.

Classification:
Currently, the CBCL classified according to WHO-EORTC classification into three main subtypes (Swerdlow 2017) which are the following:

Primary cutaneous follicle center lymphoma (PCFCL)

Primary cutaneous marginal zone lymphoma (PCMZL)

Primary cutaneous diffuse large B-cell lymphoma (PCDLBCL)

The WHO classification was updated in 2018 to include additional provisional entity, EBV-positive mucocutaneous ulcer and to recognize 2 subsets of PCMZL (Willemze 2019). Intravascular large B-cell lymphoma was also included as part of CBCL. While it has a 'cutaneous' variant (Ferreri 2004), this entity will not be discussed further in this review. It is critical to distinguish PCBCL from systemic B cell lymphomas with secondary skin involvement because the clinical behaviors, prognosis, and management differ considerably.

Clinics and pathology

Phenotype/cell stem origin

Cell Origin: CBCL, by definition, is of B-cell origin. All entities express mature B-cell phenotype. Cells are positive for a variety of B-cell markers like surface immunoglobulins (sIg), pan B-cell antigens like CD19, CD20 and/or CD79b. Each entity may be

associated with certain immunophenotypes as demonstrated below under Pathology.
Putative normal counterpart of PCMZL is post germinal center marginal zone B-lymphocytes of secondary lymphoid follicles. PCMZL is considered a type of extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT).
The PCFCL is thought to arise from mature germinal center cells B-cell while the PCDLBCL, leg-type arises from peripheral B-cells of post germinal center origin (Hoefnagel 2005).

Embryonic origin

The exact etiology and pathogenesis of CBCL is poorly understood. However, dysregulation of an antigen-driven response and infectious etiologies have been proposed in some cases. Dysregulation of germinal center B-cell proliferation might play a role in PCFCL. Infection by Borrelia burgdorferi has been implicated as etiological factor in PCMZL since DNA sequences of this organism were found in skin lesions of some PCMZL cases. However, such association may be geography- dependent since some studies have questioned this association (Ponzoni 2011).

Epidemiology

Approximately 25% of all non-Hodgkin lymphoma (NHL) cases will present at an extranodal site without systemic involvement. 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 PCBCLs make up approximately 25% of all primary cutaneous lymphomas, the remainder predominantly T-cell lymphomas (Swerdlow 2013). The incidence varies geographically with lower rates in some countries (Abeldano 2018). PCMZL represents an indolent type of CBCL comprising approximately 7% of PCL and approximately 25% of CBCL. In the revised WHO classification of lymphoma, PCMZL is considered under extranodal marginal zone lymphoma of MALT type (Swerdlow 2016, Swerdlow 2017). PCMZL usually presents in the fifth decade of life, although the disease has been diagnosed in children (Kempf 2014 Aug). Men are diagnosed approximately twice as often as women. Most cases occur in non-Hispanic Whites (Kempf 2014 Mar).
The PCFCL is the most common PCBCL, accounting for approximately 60% of such cases (Willemze 2005, Willemzie 2019). It is primarily a disease of middle aged to older patients (median age at onset is 51) and a male to female ratio of approximately 1.5:1 (Zinzani 2006, Bradford 2009, Swerdlow 2017). PCLBCL, leg type comprises 5% of all cutaneous lymphomas and 20% of all PCBCLs. The median age at presentation is in the mid to late seventies and is more common in females than in males with a male to female ratio of 1:3-4 (Swerdlow 2017). In contrast to the previous 2 entities, PCDLBCL-leg type is an aggressive disease and is associated with poor prognosis.

Clinics

Clinically, PCL is defined as non-Hodgkin's lymphoma (NHL) presenting in the skin with no evidence of extra-cutaneous disease at time of diagnosis for 6 months following initial diagnosis (Willemze 2005). This definition requires that clinicians conduct careful staging work up according to standard guidelines (Cheson 2014) to document the absence of disease elsewhere before designation as PCL. If disease is found elsewhere, like lymph nodes or other extranodal sites, then skin involvement is secondary, and the patient is designated to have stage IV lymphoma with skin involvement. As described below, none of the CBCL types has a pathognomonic appearance skin lesions and a biopsy of representative lesion is required to establish the diagnosis.

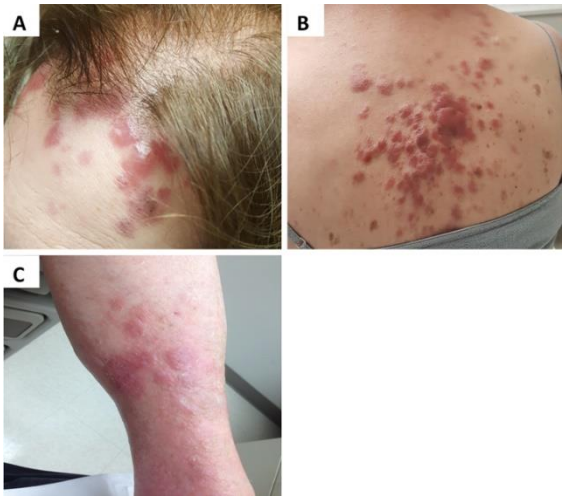

Figure 1: A: PCFCL in forehead and scalp; B: PCFCL in upper back. Skin lesions range between plaques, papules and tumor nodules which are reddish in color. C: PCDLBCL in left leg. Multiple plaque lesions

PCMZL manifests as solitary or multiple reddish, dome-shaped papules, nodules or erythematous plaques. The disease frequently presents in the trunk or extremities, and to a lesser extent in the head and neck area.
PCFCL commonly presents in the scalp and forehead (Figure 1A) but can involve the trunk and limbs. Lesions can be solitary or grouped and vary from pink papules to violaceous nodules. PCFCL that presents in the trunk (Figure 1B) used to be called reticulohistiocytoma of the dorsum or Crosti lymphoma' (Berti 1988).
PCDLBCL can be divided into 2 distinct sub-entities: PCDLBCL-leg type, and PCDLBCL-other (Paulli 2012). The leg type (LT) is commonly seen in older women and usually presents in the lower

extremities although other sites of the body can be involved.

The disease presents as rapidly growing solitary or multiple nodules or plaques (Figure 1C). The lesions are pink, red, bluish-red or violaceous in color.

Unlike PCMZL and PCFCL, DLBCL-LT is an aggressive disease where most patients will have local recurrence and/or extracutaneous progression and poor overall survival (Grange 2007). PCDLBCL-other includes T-cell/histiocyte-rich DLBCL, plasmablastic lymphoma, intravascular large B-cell lymphoma and other types that are distinct from PCDLBCL-LT.

Pathology

The diagnosis is established by performing a biopsy of the skin lesions, through histological examinations, complemented by immunophenotypic and genotypic studies (Swerdlow 2013; Kempf 2014). Histologically, three growth patterns have been recognized in PCFCL: follicular, diffuse or follicular and diffuse; the diffuse pattern is most frequent (Figure 2A).

Cells are small lymphocytes (centrocytes) with few large cells (centroblasts or immunoblasts) (Figure 2B). Immunohistochemical stain typically shows the lymphocytic infiltrate to be positive for CD20 and BCL6 but is generally negative for CD10 and BCL2 (Figure 4C-F).

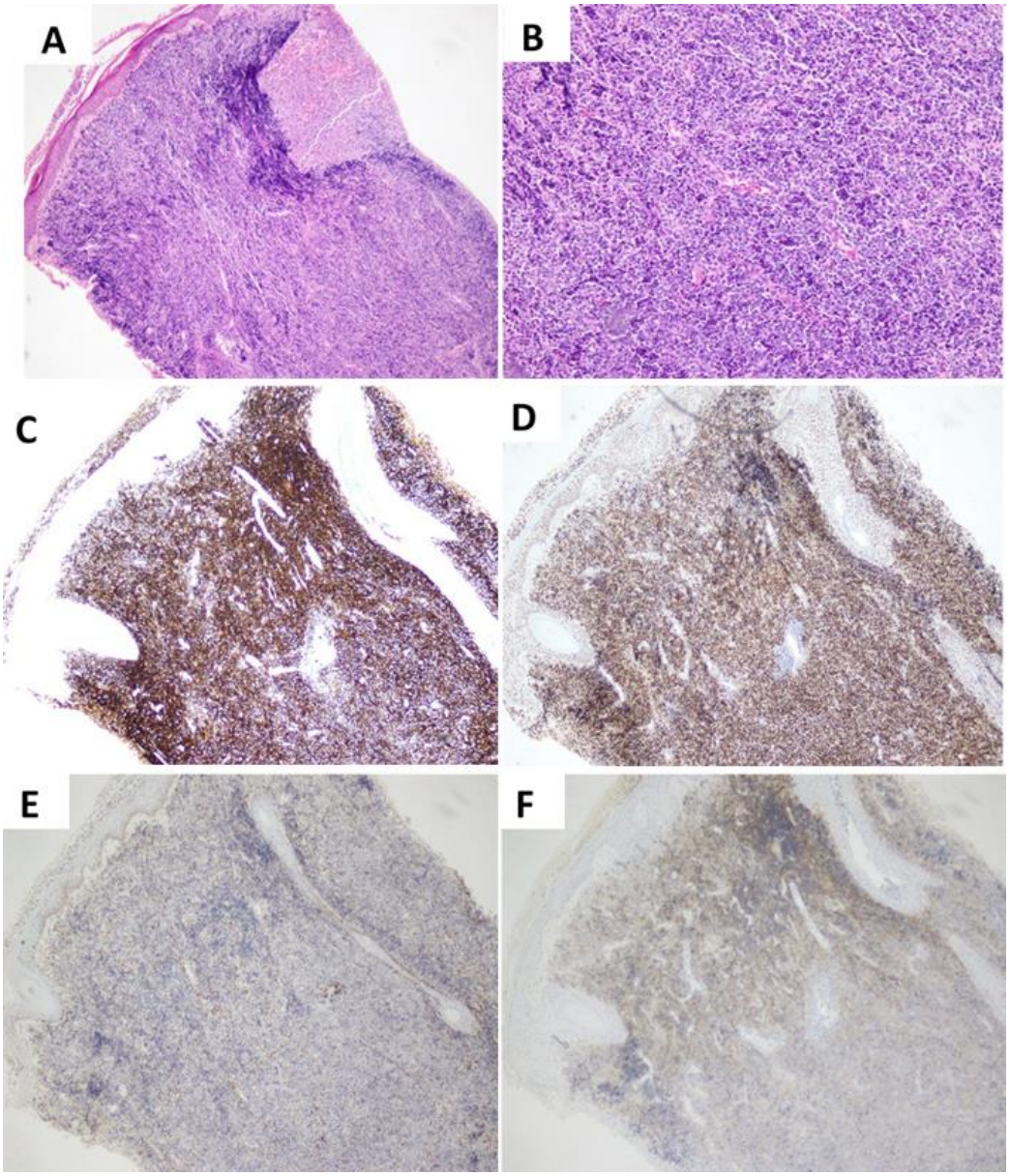

Figure 2: A, Skin biopsy of a patient with PCFCL. H & E stain; B, PCFCL, diffuse growth pattern. H & E; C, PCFCL, Immunohistochemical (IHC) stain of CD20 showing strong positive staining of lymphocytic infiltrate; D, PCFCL, IHC showing diffusely positive staining for BCL6; E, PCFCL, IHC showing positive BCL2 stain in a pattern similar to CD3 staining; 4F: PCFCL, IHC showing negative stain for CD10

Some cases may express BCL2 in a minority of malignant cells but staining is usually faint and is weaker than admixed reactive T-cells. CD10 may also be positive in some cases of follicular growth pattern but is negative in cases of diffuse growth pattern. Strong expression of BCL2 and CD10 should raise suspicion that skin involvement is secondary to a nodal follicular lymphoma rather than PCFCL. T-cell markers and CD30 are negative; however, CD3 may stain reactive T-cells which may be numerous.

Histopathologic findings of PCMZL include a nodular or diffuse nonepidermotropic infiltrate composed of small to medium-size lymphoid cells (Figure 3A). The nuclei are indented, and cells have abundant cytoplasm. Lymphoid cell infiltrate can be admixed with various number of plasma cells and reactive T-cells. Asian cases with PCMZL were reported to show tissue eosinophilia in tumor specimens, a feature not found in cases from Germany or United States (Takino 2008). Immunophenotypically, PCMZL tumor cells express B-cell markers like CD20 (Figure 3B), and CD79a. CD3 highlights reactive T-cells which sometimes are present in larger numbers than the malignant B-cells (Figure 3C). In such cases, it is important to document B-cell monoclonality which can be demonstrated using IHC techniques like in situ hybridization for immunoglobulin light chain staining. Malignant B-cells are negative for germinal center markers like CD10 and BCL6 (Figure 3D).

PCDLBCL, LT is characterized histologically by diffuse infiltrate covering the entire dermis but usually spares a thin subepidermal grenz zone and the epidermis (Figure 4A). Tumor cell infiltrate destroys adnexal structures and extends to subcutaneous tissue. As the name implies, cells are large B-cells (also referred to as centroblasts or immunoblasts) (Figure 4B). The immunophenotype of tumor cells typically is positive for CD19, CD20 (Figure 4C), CD79a, BCL2, and MUM1 (Figure 4D). Cells are usually negative for CD3 (Figure 4E), CD5, CD21, CD138, CD10, and cyclin D1. Cell proliferation is high (Figure 4F). Strong expression of BCL2 and MUM1 in PCDLBCL, LT helps distinguish this entity from diffuse type PCFCL which is negative for these 2 markers.

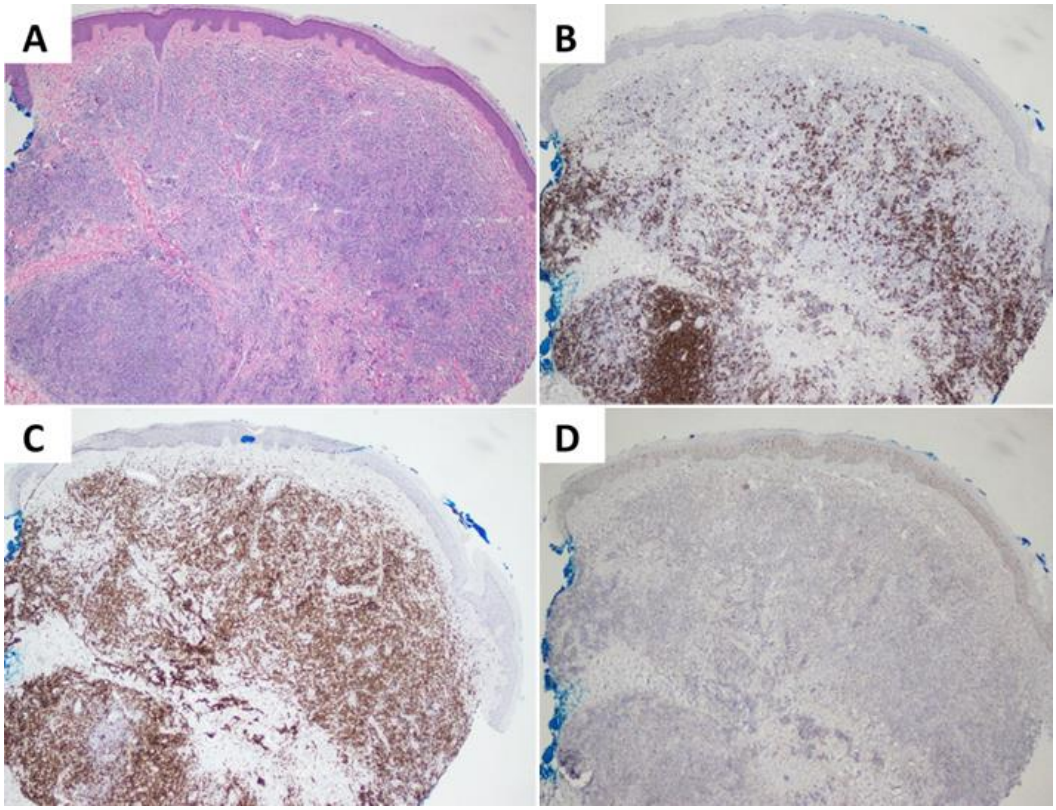

Figure 3; A: PCMZL, Skin biopsy showing deep dermal lymphocytic infiltrate; B: PCMZL. CD20 stain; C: PCMZL. CD3 stain showing abundant reactive T-cells; D: PCMZL. BCL6 stain, negative

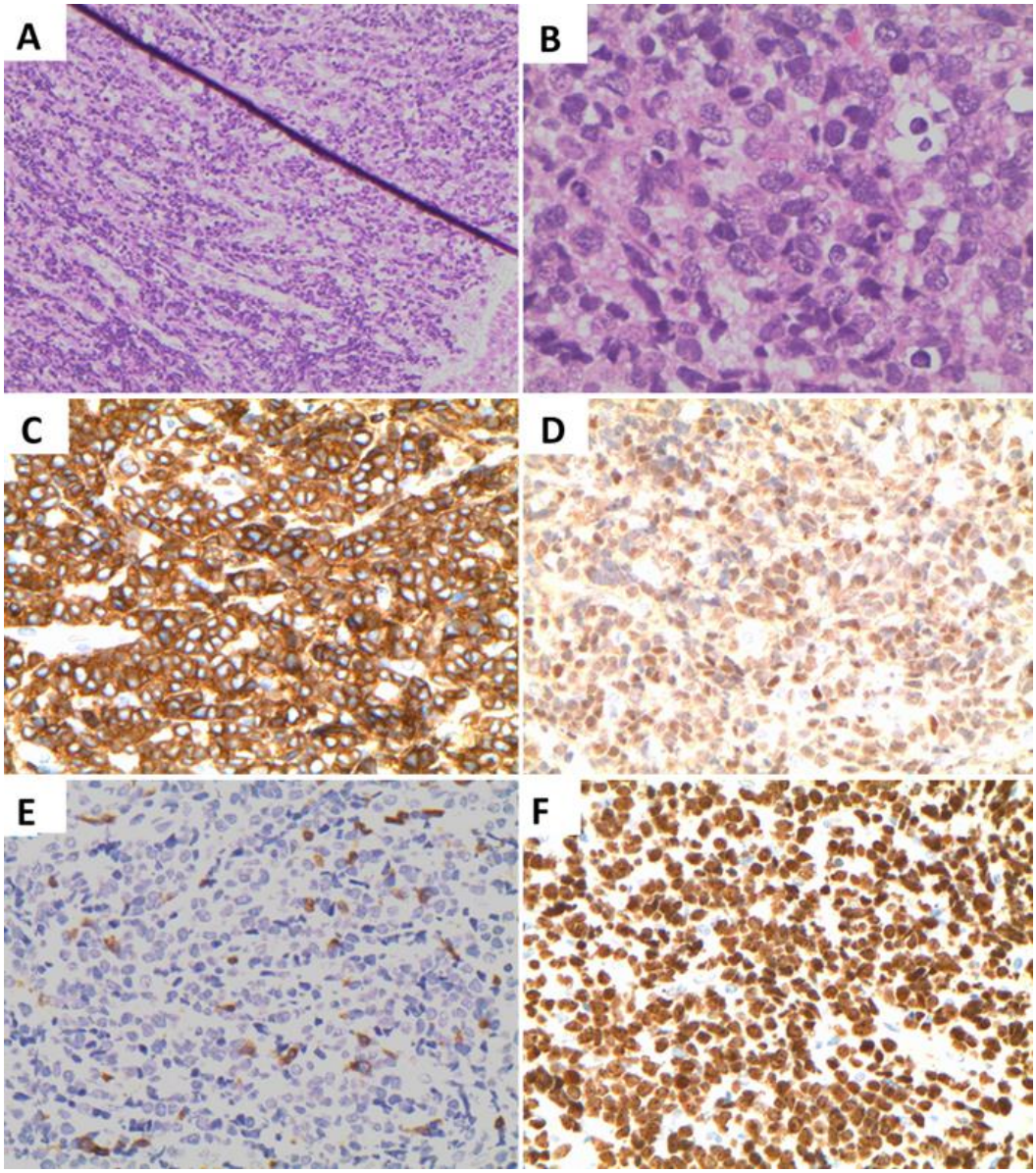

Figure 4; A: PCDLBCL, LT. H & E stain, low power showing diffuse infiltration of the dermis with sparing of the epidermis and subepidermal layers; B: PCDLBC6BL, LT. Large cells (centroblasts) with irregular vesicular nuclei and few apoptotic cells; C: PCDLBCL, LT. Strongly positive CD20 membrane stain; D: PCDLBCL, LT. MUM1 stain. Most of the large lymphoma cells are positive. E: PCDLBCL, LT. CD3 stain showing few reactive T-cells. The large cells are negative. F: PCDLBCL, LT. Ki 67. More than 90% of cells show positive nuclear stain

Treatment

The choice of therapy of CBCL relies on correct histopathologic classification and the exclusion of systemic disease. The European Organization for Research and Treatment of Cancer (EORTC) and International Society for Cutaneous Lymphoma (ISCL) have published consensus recommendations for the management of CBCL (Sneff 2008). For localized PFCL and PCMZL, the disease is managed by local therapy, such as radiation or surgical excision. Observation is appropriate for asymptomatic multifocal presentation of these categories given their indolent nature. However, for symptomatic, progressive or relapsed disease, a variety of systemic therapies are available. The simplest is anti-CD20 (rituximab) monotherapy in a standard dose/schedule.

Prognosis

Subclassification and the extent of cutaneous involvement were identified as the most relevant prognostic factors in CBCL (Zinzani 2006). PCFCL and PCMZL are indolent types of cutaneous lymphoma with excellent prognosis. The 5 year survival rate is 90-95%. However, like their nodal counterparts, they have tendency to recur in up to 40% of cases but dissemination to extracutaneous sites is rare (Chan 2016). Both PCFCL and PCMZL can transform to DLBCL with a more aggressive

behavior. The PCDLBCL, LT on the other hand is an aggressive disease with a 5 year survival of 20-50%. Mutation of the Myeloid Differentiation (MYD88) Primary Response gene (L265P) is frequently found in PCDLBCL, LT and is associated with inferior prognosis (Pham-Ledard 2014).

Genetics

Molecular study of immunoglobulin genes rearrangement is detected in all subtypes that are valuable to document clonality and differentiate PCBCL from pseudolymphomas. Until recently, cytogenetic studies had limited value in the diagnosis of **PCBCL**, since recurrent chromosomal and molecular alterations were not well characterized. Approximately 25% of **PCMZLs** demonstrate the t(14;18) (IGH/MALT1) translocation. The t(11;18) and t(3;14)orted in 7% and 10% of PCMZLs, respectively, whereas the t(1;14) translocation has not been identified in PCMZL (Abdul-Wahab, 2014). MYD88 mutations have been found in 50% of non-class-switched cases of PCMZL, but not in class-switched cases (Wobser 2017). Recurrent mutations in FAS were found in 24 of 38 (63%) patients with PCMZL (Maurus 2018).The overexpression of microRNAs MIR 155 and MIR150 is another molecular feature unique to PCMZL, and this feature may also predict longer progression-free survival.

On the contrary, the **PCDLBCL, LT** shows many genetic similarities with diffuse large B cell lymphomas arising at other sites. Interphase FISH analysis frequently shows translocation involving MYC/8q24, BCL6/3q27 and IGH/14q32 genes in PCDLBCL, LT but not in patients with a PCFCL (Hallermann 2004 July). Comparative genomic hybridization (CGH), using microarrays and subsequently confirmed by FISH analysis showed that most of PCDLBCL, LT cases had chromosomal aberrations including gains in chromosomes 1, 2, 3, 7, and 12, losses in 6q, 13, 14, and 18q (Dijkman 2006). In contrast, PCFCL had fewer imbalances and lacked translocations affecting the IGH locus (Hallermann June 2004). Most PCFCL do not exhibit t(14;18)(q32;q21) that determines BCL2/IGH rearrangement and features of nodal follicular lymphoma (Abdul-Wahab 2014). The most recurrent alterations in PCFCL were high-level DNA amplifications at 2p16.1 (63%) and deletion of chromosome 14q32.33 (68%). FISH analysis confirmed REL amplification in patients with gains at 2p16.1. The same study showed that PCDLBCL, LT have a high-level DNA amplification of 18q21.31-q21.33 (67%), including the BCL2 and MALT1 genes as confirmed by FISH. This may explain the strong BCL2 expression in these cases although lacking t(14;18) (BCL2/IGH) translocation. Recurrent homozygous DNA deletions in region 9p21.3 which contains CDKN2A, CDKN2B and NSG-x genes are found in 67% of PCDLBCL, LT cases but not in any of the PCFCL patients (Belaud-Rotureau 2008). In addition, some PCDLBCL, LT (17%) had a complete hypermethylation of CDKN2A gene promoter. In contrast, deletions of chromosome 9p21.3 containing the CDKN2A and CDKN2B gene loci, and MYD88 mutations are not or only rarely found in PCFCL. In conclusion, these studies clearly demonstrate distinct chromosomal and genomic aberrations in PCFCL and PCDLBCL, LT which provides further support for their categorization in the WHO-EORTC classification as separate entities. Finally, loss of chromosome 9p21.3 might prove to be an important prognostic marker in PCDLBCL, LT associated with inferior outcome (Sneff 2009) although confirmation on a larger group of patients is required. PCFCL shows the gene expression profile of germinal center-like large B cell lymphomas, and often shows amplifications of the REL gene (Hoefnagel 2005). Deletion of chromosome 14q32.33 has been also reported in PCFCL (Dijkman 2006).

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