



# Lymphoproliferative Disorders Mimicking Rheumatologic Disease: A Clinical Reasoning Perspective

Jeffrey Z. Shen<sup>1,8</sup> · Eric D. Carlsen<sup>2,3</sup> · Luis F. Carrillo<sup>2</sup> · Dahima Cintron<sup>4</sup> · Ben Kellogg<sup>1</sup> · Jamie Lim<sup>1</sup> · Matilda Nicholas<sup>1,5</sup> · Elijah Lackey<sup>1,6</sup> · John Dasher<sup>7</sup>

Received: 28 June 2025 / Accepted: 15 August 2025

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## Abstract

**Purpose of Review** Lymphoproliferative disorders encompass a variety of hematologic diseases that are increasingly recognized. Patients with these conditions may present to rheumatologists or other specialists prior to a hematologist-oncologist. This review is intended to highlight key distinguishing features between rheumatologic conditions and lymphoproliferative disorders, and suggest diagnostic approaches.

**Recent Findings** Recent studies have shown that lymphoproliferative diseases can mimic rheumatologic disease, and differentiation is complex. Advances in imaging techniques such as FDG PET/CT, and improved understanding of lymph node histopathology have improved diagnostic accuracy. Attention to specific details such as the pattern of organ involvement, the presence or absence of key clinical features, and the consistency of treatment response can assist in accurate diagnosis.

**Summary** Our review highlights the importance of considering lymphoproliferative disorders in the differential diagnosis of rheumatologic disease. A thorough clinical history, targeted biopsies, and collaboration with hematopathologists are essential for timely diagnosis and care. Future research should concentrate on identifying biomarkers and formulating diagnostic algorithms to aid in differentiating lymphoproliferative disorders from autoimmune diseases, which will ultimately improve patient outcomes.

**Keywords** Lymphoma · Lymphadenopathy · Lupus · Diagnosis · Mimics

## Introduction

Lymphoproliferative disorders (LPDs) comprise a group of malignant and benign hematologic conditions characterized by proliferation of abnormal B cells, T cells, and/or natural killer (NK) cells [1]. Patients with these conditions may initially present to rheumatologists, or general internists, rather than hematologists due to overlapping clinical features as well as shared laboratory, imaging, and histopathologic findings with autoimmune diseases [2]. LPDs may exhibit response to corticosteroids in a fashion similar to rheumatologic disease. However, this response eventually wanes, potentially resulting in corticosteroid toxicities, permanent end-organ damage or, in some cases, transformation of an indolent LPD into an aggressive hematologic malignancy [3, 4]. Additionally, patients may have co-existing rheumatologic disease with LPD, such as Rheumatoid Arthritis (RA) with Large Granular Lymphocytic Leukemia (LGLL) or Sjogren's Disease with Mucosal-Associated Lymphoid Tissue Lymphoma (MALT-L) [5–7]. Rarely, patients with

✉ Jeffrey Z. Shen  
jzs4@duke.edu

<sup>1</sup> Duke Department of Medicine, Division of Rheumatology and Immunology, NCDurham, USA

<sup>2</sup> Duke Department of Pathology, Division of Hematopathology, NCDurham, USA

<sup>3</sup> Duke Cancer Institute, NCDurham, USA

<sup>4</sup> Duke Department of Medicine and Pediatrics, Division of Rheumatology, NCDurham, USA

<sup>5</sup> Duke Department of Dermatology, NCDurham, USA

<sup>6</sup> Duke Department of Neurology, Division of Neuroimmunology, NCDurham, USA

<sup>7</sup> Birmingham Department of Medicine, Division of Hematology and Oncology, University of Alabama, ALTuscaloosa, USA

<sup>8</sup> Duke Medicine Cir Clinic 1J, 27710 Durham, NC, USA

adult-onset primary immunodeficiencies can present as lymphoproliferative disorders with rheumatologic features. Understanding of LPDs has improved significantly recently with the advent of next generation sequencing [8].

This review examines clinical scenarios where adult rheumatologists may face LPDs masquerading as autoimmune conditions, offer literature-based guidance in these situations, and identify knowledge gaps where future studies can improve diagnosis. We compare clinical, exam, laboratory, imaging, and pathologic features that overlap between LPDs and rheumatologic disease, and offer guidance on when to pursue hematology referral.

## Investigating the Etiology of Lymphadenopathy

### Distribution of Lymphadenopathy

Rheumatologists commonly encounter diffuse lymphadenopathy (LAD), defined here as LAD in at least two separate non-contiguous regions. Fever and B symptoms (chills, night sweats, fatigue, unintentional weight loss, malaise) can occur commonly in both malignant and reactive LAD from autoimmune disease or infection [9]. While diffuse LAD is common in certain rheumatic diseases such as Systemic Lupus Erythematosus (SLE) it is absent in others such as vasculitis or seronegative arthritis (Supplemental Table 1) [10–33]. A study of 38 SLE patients presenting with LAD identified no microbial RNA or DNA by shotgun metagenomics, suggesting that the LAD may be directly related to SLE rather than an undiagnosed infection [34]. Further investigation is required to understand whether this finding extends to other rheumatologic causes of LAD.

In patients with diffuse LAD, fever of unknown origin (FUO) decreases the likelihood of sarcoidosis and IgG4-Related Disease (IgG4-RD), but is compatible with SLE or Sjogren's (Supplemental Table 1) [15, 35, 36]. Lymphomas, leukemias, and Multicentric Castleman's Disease (MCD) are important LPDs to consider for patients who present with FUO and diffuse LAD. Non-Hodgkin Lymphomas (NHL) account for approximately 90% of all lymphomas [37]. The most common NHLs are Diffuse Large B-cell Lymphoma (DLBCL, 30% of cases), Follicular Lymphoma (FL, 20% of cases), and Marginal Zone Lymphoma (MZL, 10% of cases) [37]. While DLBCL is an aggressive subtype, both FL and MZL typically follow a more indolent course that can mimic rheumatologic disease [38, 39]. LPDs can frequently present with positive autoantibodies (Table 1).

Positive anti-nuclear antibody (ANA) results on commercial laboratory tests can be seen in up to 30% of patients with NHL and 5% of those with Hodgkin lymphoma (HL)

**Table 1** Overlapping laboratory findings between rheumatologic and lymphoproliferative diseases

Laboratory Result	Most Common Rheumatologic Condition	Most Common LPD
ANA+	CTDs	NHLs MCD
Specific ENA (dsDNA+, SSA+)	CTDs	DLBCL MZL (MALT-L) MCD
RF+	RA CTDs ANCA vasculitis CryoV	MZL (MALT-L) CLL LPD causes of CryoV
CCP+	RA	DLBCL
ANCA+	ANCA vasculitis	NHLs CLL MDS*
DAT+	CTDs (SLE)	CLL AITCL
Elevated serum IgG4	IgG4-RD HUVS ANCA vasculitis (rare) Sjogren's (rare)	DLBCL MZL MCD Histiocytosis*
Elevated serum ACE	Sarcoidosis GPA EGPA Seropositive RA (nodules)	HL LYG

In this table, we list the LPDs that most commonly cause the corresponding false positive laboratory result. LPDs may also rarely cause spurious polyspecific autoantibody reactivity, in which multiple unrelated autoantibodies are present without their corresponding pathologies

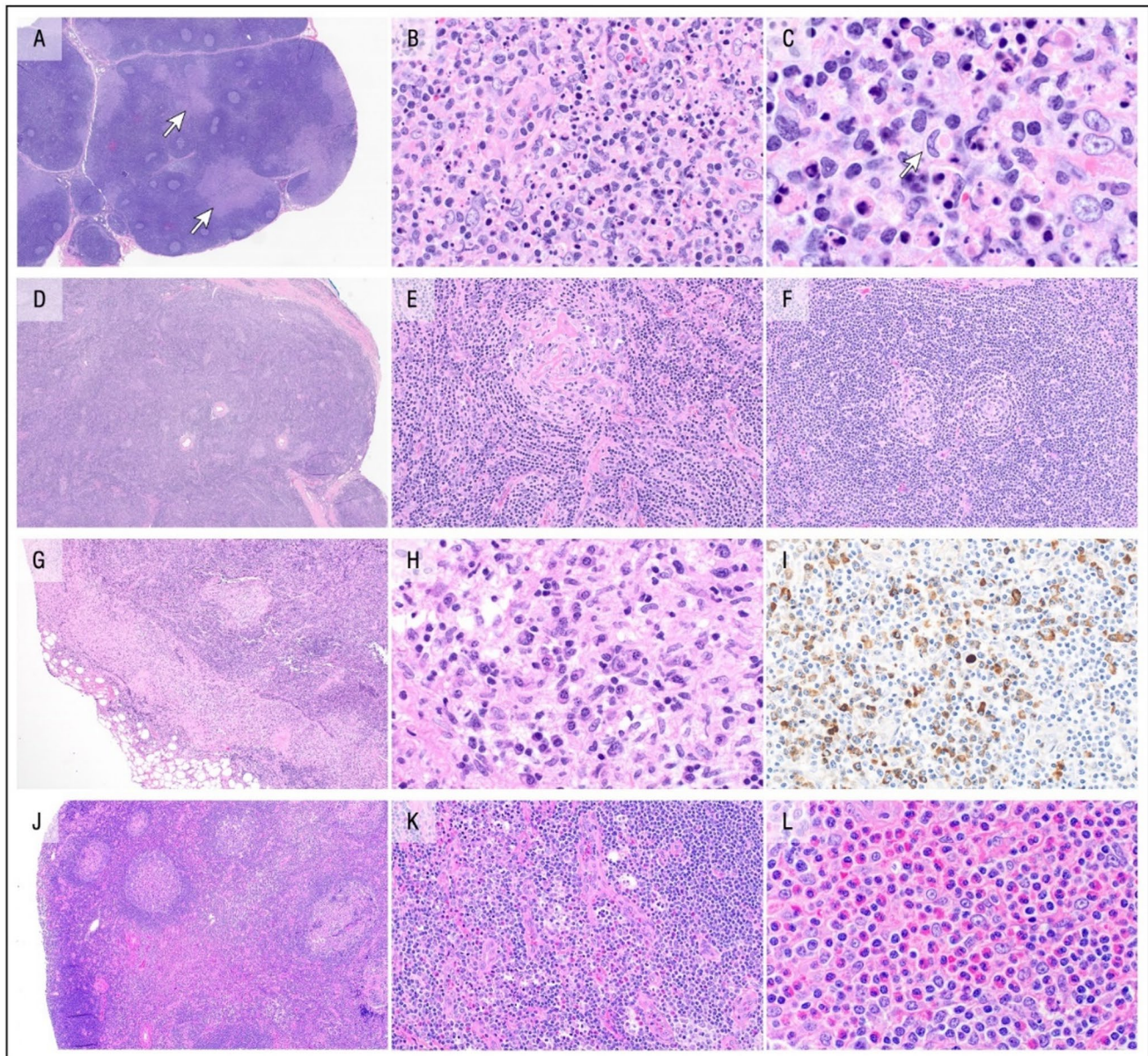
*CTDs* Rheumatologic connective tissue diseases, which we herein define as SLE, Sjogren's, Scleroderma, Myositis, Mixed connective tissue disease, and other overlap syndromes, *ENA* Extractable nuclear antigen including antibodies to dsDNA, SSA, SSB, Sm, RNP, *CryoV* Cryoglobulinemic vasculitis, *CLL* Chronic lymphocytic leukemia (exists on a spectrum with small lymphocytic lymphoma, SLL), *DAT* Direct Antiglobulin Testing, *HUVS* Hypocomplementemic urticarial vasculitis, *MALT-L* Mucosal Associated Lymphoid Tissue Lymphoma (a subtype of extranodal MZL), *LYG* Lymphomatoid granulomatosis, *GPA* Granulomatosis with polyangiitis, *EGPA* Eosinophilic granulomatosis with polyangiitis, *MDS* Myelodysplastic syndrome, *AITCL* Angioimmunoblastic T cell Lymphoma. \*These are technically not LPDs, but can have lymphoproliferative features

[40]. Specific autoantibodies to extractable nuclear antigens (ENA) such as anti-dsDNA are far less common, but the occurrence is difficult to approximate due to the variability of testing methods for ENA [41, 42]. Positive ANCA or positive testing for lupus anticoagulant can also be seen [43, 44]. In one large study, clinical symptoms suggestive of rheumatologic disease were seen in 30% of ANA positive NHL patients [45]. The prevalence of true autoimmunity in patients with DLBCL is about 20%, with RA being the most common rheumatologic disease [46].

Rheumatologic diseases such as SLE may also cause isolated LAD, localized to regions such as the cervical, mediastinal, or retroperitoneal lymph nodes (LN). Excisional LN

biopsy may be necessary to differentiate rheumatologic diseases from LPD mimics such as Kikuchi's Disease (Fig. 1, A-C) and MCD (Fig. 1, D-F) in these contexts.

Classical HL (cHL, 95% of HL) presents initially as unilateral cervical LAD and metastasizes vertically rather than horizontally to the next contiguous LN station



**Fig. 1** Lymph node histologic patterns. Panels (A-C) represent histiocytic necrotizing lymphadenitis, a pattern that is seen in both Kikuchi-Fujimoto disease and lupus lymphadenitis. Panel (A) shows intact lymph node architecture with wedge-shaped areas of paracortical necrosis (white arrows) (Hematoxylin and eosin stain; 2x objective). Panel (B) shows higher magnification of necrotic areas containing abundant apoptotic debris and a conspicuous absence of neutrophils (20x objective). Panel (C) demonstrates a higher magnification of the necrotic areas, revealing apoptotic debris and crescentic histiocytes (white arrow) (50x objective). Panels (D-F) illustrate features of hyaline-vascular Castleman disease. Panel (D) shows a low-magnification view of regressed germinal centers and increased interfollicular vessels (Hematoxylin and eosin stain; 2x objective). Panel (E) shows an involuted follicle with hyalinized vessels and concentric layering of the

mantle zone (“onion skinning”) (10x objective). Panel (F) reveals multiple germinal centers within a single follicle (so-called “twinning”) (4x objective). Panels G-I represent IgG4-related lymphadenopathy. Panel G shows a lymph node with capsular sclerosis (Hematoxylin and eosin stain; 2x objective). Panel (H) demonstrates numerous plasma cells within the sclerotic areas (20x objective). The plasma cells in panel I reveal an increased number of IgG4-positive plasma cells (representing >40% of IgG (+) cells) (IgG4 immunohistochemical stain) (10x objective). Panels (J-L) illustrate features of Kimura disease. Panel (J) shows a lymph node with follicular hyperplasia (Hematoxylin and eosin stain; 2x objective). Panel K demonstrates an eosinophilic infiltrate and vascular proliferation (10x objective). Panel L shows an eosinophilic microabscess in the paracortex (40x objective)

(mediastinum) unlike other LPDs such as NHLs and nodular lymphocyte predominant HL (NLPHL, 5% of HL) [47]. Bilateral cervical LAD can occur in late stage cHL but represents widespread metastatic disease [47]. Large B-cell Lymphoma with IRF4 rearrangement is a prominent DLBCL subtype that commonly presents as isolated cervical LAD. It may mimic a reactive follicular pattern on biopsy and can also involve Waldeyer's ring as a localized mass [48–50]. Table 2 highlights key features to differentiating LPDs from rheumatologic disorders presenting as isolated cervical LAD.

Supplemental Tables 2 and 3 are intended to guide clinicians on when to pursue workup for LPDs in patients presenting with isolated mediastinal LAD or retroperitoneal fibrosis (RPF) [12, 49, 51–69].

### Exam and Imaging

Physical exam can detect LAD, but may not be sufficient to distinguish causes with adequate certainty. Imaging studies may assist with diagnostic evaluation, but LN size > 1 cm can be caused by rheumatologic disease in addition to LPDs [10, 11, 14, 34, 70, 71]. Sarcoidosis frequently causes intrathoracic LAD larger than 2 cm in diameter [72, 73]. 18 F-fluorodeoxyglucose Positron Emission Tomography with Computed Tomography (18 F-FDG PET/CT) has >97% sensitivity for aggressive lymphomas and has superiority over standard CT [74–76]. LNs in aggressive lymphoma

subtypes are known to be more FDG avid on PET/CT than reactive LNs, but this is less true for indolent lymphomas [77, 78]. An intense and asymmetric Pattern of FDG uptake in LNs has been shown in one study to suggest HIV-associated lymphoma with 90% accuracy over reactive LAD in HIV [79]. Future studies should assess and compare the size, symmetry, and FDG uptake of LAD in different autoimmune diseases.

### Lymph Node Biopsy

In patients with diffuse LAD, LN biopsy (core or excisional) is the best test to differentiate reactive LAD from LPDs [80]. Patterns of LAD in SLE, RA, and Sjogren's are generally nonspecific. LNs affected by IgG4-RD may exhibit a variety of histologic patterns, but all display an increase in total IgG4 positive plasma cells and a high IgG4/IgG plasma cell ratio (Fig. 1, G-I). Absence of reported immunohistochemical stains for IgG4 suggests that the pathologist has not yet assessed for IgG4-RD. Sarcoidosis classically causes "naked" (i.e. minimal background lymphocytes) well-formed non-necrotizing granulomas, but their abundance is highly variable. LPDs and other mimics, in contrast, can cause poorly formed granulomas or an infiltrate that resembles granulomatous inflammation, but the presence of well-formed non-necrotizing granulomas is less common (Table 3) [81–89]. Future studies should compare the patterns of granulomatous inflammation across

**Table 2** Differential diagnosis of isolated cervical lymphadenopathy, using only five criteria

Conditions	H&P	Lab Findings			Path
	Painful LAD	ANA+	Persistent Eosinophilia	High IgG4	Granulomas
<b>Rheumatologic</b>					
SLE					
Sjogren's					
Sarcoidosis*					
IgG4-RD*					
<b>LPD</b>					
Kikuchi					
DLBCL					
Hodgkin's					
AITCL					
<b>Other</b>					
RDD					
Kimura's					

This table highlights key features distinguishing rheumatologic conditions from lymphoproliferative mimics for patients presenting with isolated cervical lymphadenopathy. Other common etiologies (infections, medication reactions, immunodeficiencies) are important to consider, but not addressed here. RDD Rosai-Dorfman Disease. \*It is very rare for sarcoidosis or IgG4-RD to cause cervical LAD without additional lymph node involvement.

**Table 3** Overlapping pathologic findings between rheumatologic and lymphoproliferative diseases

Pathology	Lymph Node	Skin	Liver	Kidney	Sural Nerve	Muscle	Brain
<b>Non-Caseating Granulomas</b>							
HL							
DLBCL							
AITCL							
CTCL							
ENKTCL							
CLL							
<b>Necrotizing Vasculitis</b>							
PCNSL							
LYG*							
IVL							
HCL							
CLL							
WM (CryoV)							
<b>Granulomatous Vasculitis</b>							
HL							
DLBCL							
CTCL							
AITCL							

This table illustrates how LPDs can be confused for rheumatologic disease even after tissue biopsy is performed. Although LPDs can demonstrate granulomas or vasculitis on biopsy, these may not be found in every organ. A green box indicates that the LPD has been identified in the literature to cause the corresponding finding in the organ indicated by the column. *CTCL* Cutaneous T-Cell Lymphoma, *ENKTCL* Extranodal NK/T Cell Lymphoma, *PCNSL* Primary CNS Lymphoma (usually DLBCL subtype), *IVL* Intravascular Lymphoma (usually DLBCL subtype), *HCL* Hairy Cell Leukemia, *WM* Waldenstrom’s macroglobulinemia (which causes type I CryoV). \*LYG can be confused on biopsy as non-caseating granulomas, necrotizing granulomas, or granulomatous vasculitis

sarcoidosis, atypical infections, LPDs, rheumatologic conditions, and immunodeficiencies.

### Techniques Used in the Pathologic Diagnosis of Lymphoproliferative Disorders

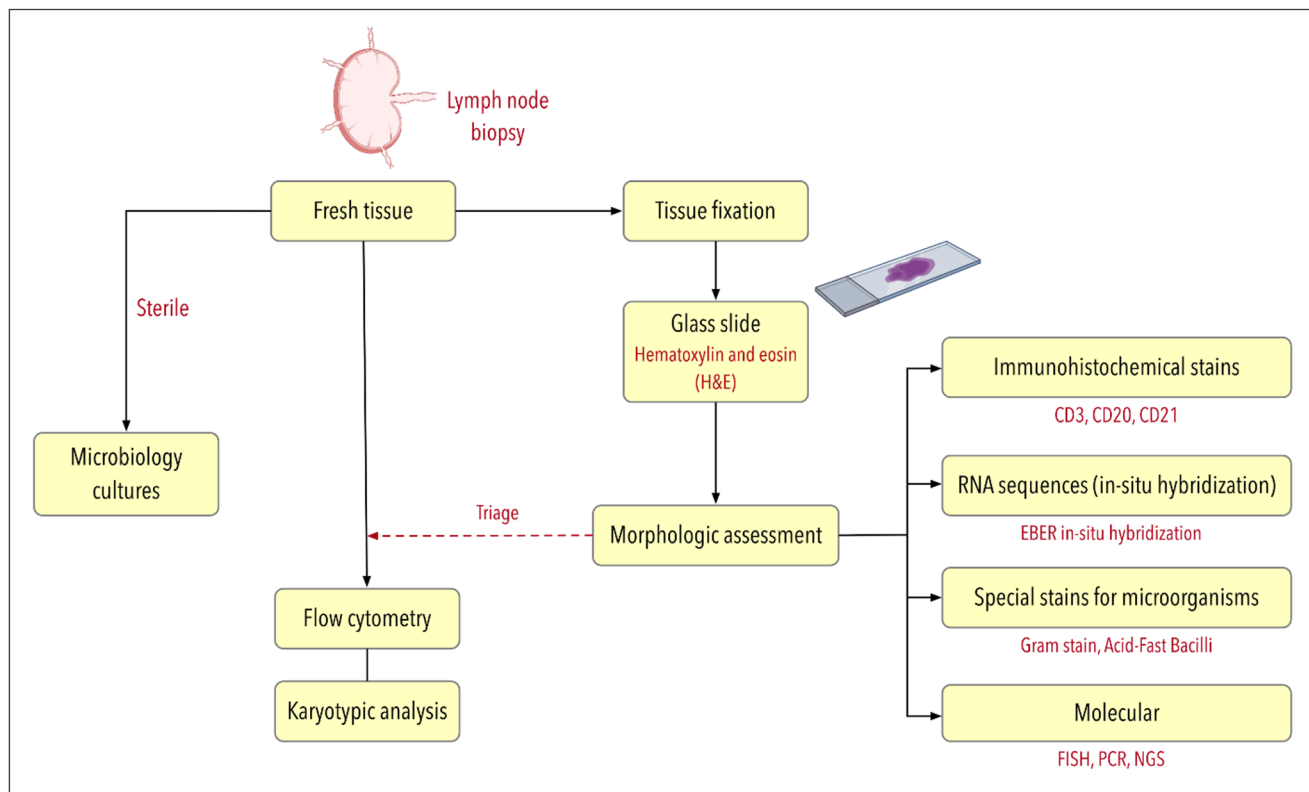
#### Histologic/cytologic Assessment

Hematoxylin and eosin-stained sections (H&Es) are routinely produced in the histologic assessment of LN and bone marrow core biopsies and serve as the cornerstone for pathologic evaluation of LPDs and reactive mimics [90]. Biopsy material is fixed in formalin and embedded in paraffin (FFPE) blocks so that very thin sections can be cut and stained. After reviewing the patient’s clinical history, pathologists formulate differential diagnoses based on initial H&E findings [91]. Differential diagnostic considerations are then resolved with the aid of additional stains for specific antigens (interrogated by immunohistochemistry), for RNA sequences (via in situ hybridization), or for microorganisms (Gram stain, Acid-Fast Bacilli, etc.). *Material allocated for H&Es and other histologic stains cannot be used to perform microbial culture, flow cytometry, or karyotype* which

can limit retrospective sample analysis (see below). However, many centers are validated to perform fluorescence in situ hybridization (FISH) and PCR-based assays on FFPE (Fig. 2).

Fresh material is allocated for microbiology cultures (collected under sterile conditions in the operating room), flow cytometric analysis, or karyotyping, as clinically indicated. The remaining tissue is formalin-fixed, paraffin-embedded, and hematoxylin and eosin-stained sections are prepared for morphologic evaluation and further triage. Ancillary studies (immunohistochemical stains, RNA sequences, special stains, or molecular studies) are performed based on initial histologic findings. EBER denotes Epstein-Barr encoding region, FISH fluorescence in situ hybridization, PCR polymerase chain reaction, and NGS next-generation sequencing.

Bone marrow aspirate material is also routinely smeared onto slides and stained with a Wright preparation for visualization under high magnification, offering superior sensitivity for detecting cytologic atypia (e.g. dysplastic changes in hematopoietic precursors) and cell identification (e.g. distinguishing blasts from more mature myeloid or lymphoid cells) [92]. Cytochemical stains can be performed on bone marrow aspirate (iron stain, myeloperoxidase, nonspecific



**Fig. 2** Lymph node tissue processing

esterase) but the breadth of testing is limited compared to that seen in immunohistochemistry (See Supplemental Fig. 1).

### Flow Cytometry

Flow cytometry uses laser optics to measure some physical properties of cells such as size and interior complexity. Antigen expression can also be interrogated using antibodies conjugated to various fluorescent dyes called fluorophores [93]. Flow cytometry is routinely performed on LNs, bone marrow aspirates, and peripheral blood but can also be performed on samples from other anatomic sites after tissue disruption. This assay is best suited for identifying phenotypic changes on hemolymphoid cell populations and plays a critical role in the diagnosis of B-cell, T-cell, and NK cell neoplasms [94].

### Pathologic Evaluation of Lymph Node

Pathologic assessment of LNs is usually via fine needle aspiration (FNA), core biopsy, or excisional biopsy. Generally, isolated FNA is insufficient or suboptimal in the evaluation for lymphoproliferative disorders because an assessment of LN architecture cannot be performed [95]. Core biopsy, particularly when paired with flow cytometric analysis, is

superior to FNA for the diagnosis of histologically distinct LPDs such as classical HL, DLBCL, or anaplastic large cell lymphoma (ALCL). However, core biopsy may not be able to distinguish subtle or morphologically variable LPDs such as angioimmunoblastic T-cell lymphoma (AITCL) and MZL, from reactive LAD [96].

If the cause of LAD is unknown or an LPD is suspected, fresh biopsy material should be allocated for flow cytometric studies. Fresh material may also be set aside for culture or to perform karyotypic analysis, a practice utilized at some but not all centers, which is useful for prognostication in many hematologic malignancies [97, 98]. Neither of these specialized studies can be performed on FFPE tissue. Thus, communication with the gross pathology laboratory at the time of biopsy is critical so that material can be triaged appropriately.

LN architectural preservation is a critical first low-power assessment in the histologic evaluation of lymphoproliferative disorders. Normal and reactive LNs are organized into distinct compartments, which are usually well-demarcated and enriched for certain, predictable cell types [90]. On cross-sections of benign LNs of sufficient size, it should be relatively easy to identify areas with B-cell-rich follicles and parafollicular accumulations of T-cells in the cortex and plasma cell and histiocyte-rich cords surrounding endothelial sinuses in the medulla. The zonation of these distinct

compartments may become blurred in some reactive conditions (particularly certain viral infections including HIV and EBV), but architectural preservation is generally a good indicator of a benign etiology (Supplemental Fig. 2, A-C) [99]. In contrast, architectural effacement is often seen in high grade lymphomas (Supplemental Fig. 2, D-F). Architectural effacement may be oblitative (i.e. normal LN structures are completely destroyed) or may be more subtle (i.e. abnormal follicles growing in compartments where follicles should not be present). Evaluation for architectural preservation is often aided by additional immunohistochemical stains [100].

In reactive LNs, architecture is typically preserved or subtly distorted, and certain histologic features may be accentuated. For example, reactive follicular hyperplasia (Supplemental Fig. 2A), a common pattern in reactive LAD, exhibits an increase in the size and abundance of LN follicles. This pattern of histologic change in LNs is common in autoimmune disease and various localized or systemic infections [101].

If architectural effacement is identified at low-power (Supplemental Fig. 2D), strategic examination at high-power takes place to better appreciate cell morphology. This high-power assessment is often helpful for identifying diagnostic features, such as “hallmark” cells in ALCL, Reed-Sternberg cells in classic HL, and centroblasts or immunoblasts in DLBCL. This stepwise approach to histologic evaluation narrows the differential diagnosis before confirmatory immunohistochemical (Supplemental Fig. 2E) and/or flow cytometric evaluation (Supplemental Fig. 2F) is performed [102].

### Cytopenia: Distinguishing Rheumatologic from Lymphoproliferative Causes

Cytopenias can occur frequently in rheumatology patients with CTDs (especially SLE). This is primarily due to active autoimmune disease but can also result from medication side effects or infectious complications of immunosuppression [103–105]. A retrospective single-center study on patients with rheumatologic disorders referred to hematology for unexplained cytopenia found that clonal hematologic processes were common (22%) but neither the degree of cytopenia for any cell line, nor the presence of single versus multiple cytopenia could help differentiate cytopenia reactive to the rheumatologic disease from the development of hematologic disorders including LPDs [106].

We reviewed the literature on each cytopenia to assess whether features such as severity, time course, size of the respective cell line, smear, type of cytopenia, other laboratory findings, or co-occurrence with other clinical features

(i.e. fever, LAD, or splenomegaly) could reliably distinguish LPD from rheumatologic disease. In patients with persistent or progressive unexplained cytopenia with diffuse LAD and/or splenomegaly, LPDs should be considered, especially if other red flags findings are present (See Supplemental Table 4) [5, 103, 106–122].

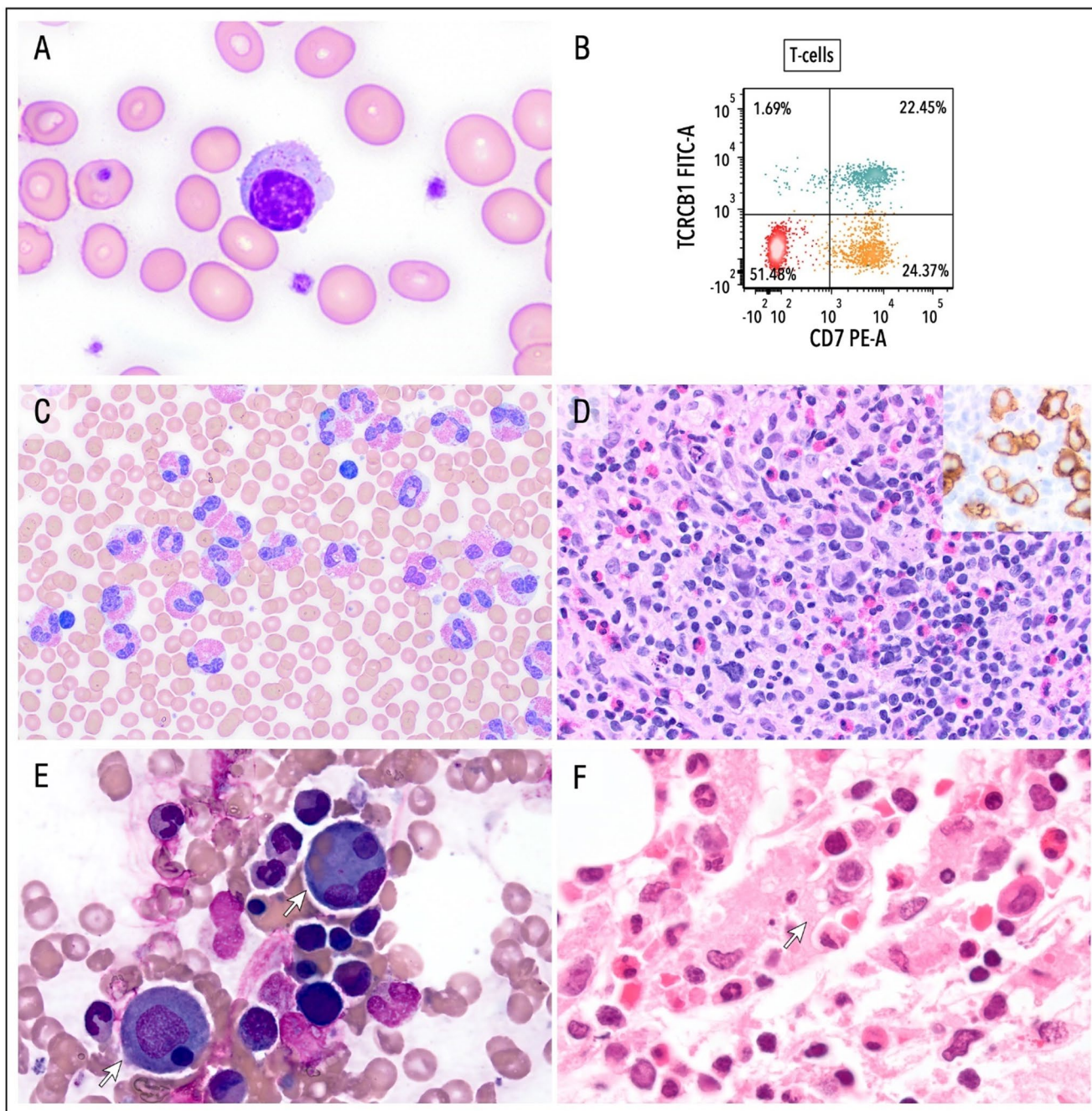
Although pancytopenia can occur in SLE, MAS, and very rarely in sarcoidosis, it is also commonly caused by LPDs [123, 124]. In a study on adults with new onset pancytopenia, 64% were due to clonal hematopoietic disorders [23]. Myeloid processes accounted for the majority of these cases, but some patients were found to have lymphoid neoplasms such as NHL, hairy cell leukemia (HCL), and precursor B acute lymphoblastic leukemia (ALL) [23].

In patients with rheumatologic diseases, the presence and degree of cytopenia often correlates with overall disease activity and is responsive to immunosuppressive therapy [125]. In practice, medication nonadherence is a major cause of cytopenia in patients with SLE. In patients with other rheumatologic disorders, medication toxicity can also be a common cause of cytopenia. However, evaluation of whether the cytopenia is caused by medication nonadherence or toxicity often takes weeks, so symptomatic patients should undergo concurrent evaluation for other etiologies. Consideration for referral to hematology is recommended when cytopenia occurs with any of the red flags mentioned above, when it is accompanied by clonal lymphocyte populations (Fig. 3, A-B), or when it is severe/progressive without a clear explanation [106].

### Hemophagocytic Lymphohistiocytosis: Distinguishing Rheumatologic from Lymphoproliferative Causes

Hemophagocytic Lymphohistiocytosis (HLH) is a severe life-threatening, hyper-inflammatory syndrome that is classified as primary or secondary by etiology. Primary HLH, also known as familial HLH, is a rare genetic condition that most commonly presents in children caused by mutations in enzymes that are crucial for the normal cytotoxic function of lymphocytes and/or NK cells [126, 127]. Although there are subtle differences in the fundamental pathophysiology, secondary HLH is an acquired condition that has similar findings to primary HLH [127, 128].

Rheumatologic patients may develop a process called macrophage activation syndrome (MAS) which clinically resembles secondary HLH. SLE and Adult-Onset Still's Disease (AoSD) are the two most common autoimmune causes of HLH/MAS in adults [127]. However, autoimmune diseases are not the most common cause of HLH. In a review of 661 HLH patients, infections and malignancies



**Fig. 3** Morphologic findings in peripheral blood and bone marrow. Panels (A–B) represent a T-cell large granular lymphocyte leukemia (T-LGL) in a patient with Felty syndrome. Panel (A) shows a large granular lymphocyte in a peripheral blood smear (Wright-Giemsa stain; 100x objective). The flow cytometric analysis in panel (B) shows an abnormal T-cell population (red) with loss of CD7 expression and TCRB1 restriction, consistent with a clonal T-cell process. Panels (C–D) illustrate hypereosinophilia in a patient with Hodgkin lymphoma. Panel (C) shows increased eosinophils in a peripheral blood smear (Wright-Giemsa stain; 50x objective). Panel (D) demonstrates a lymph node biopsy with frequent eosinophils and scattered large-sized cells

(Reed-Sternberg cells), consistent with Classic Hodgkin lymphoma (Hematoxylin and eosin stain; 20x objective). **D, inset:** CD30 immunohistochemical stain highlighting Reed-Sternberg cells (40x objective). Panels (E–F) represent Hemophagocytic Lymphohistiocytosis (HLH) in a patient with aggressive NK-cell leukemia. Panel (E) shows large histiocytes containing intact intracellular RBCs and lymphocytes in a bone marrow aspirate (white arrow) (Wright-Giemsa stain; 50x objective). Panel (F) shows hemophagocytic histiocytes in the core biopsy, with engulfed erythrocytes and granulocytic precursors evident within the histiocyte cytoplasm (white arrow) (Hematoxylin and eosin stain; 40x objective)

accounted for more than 70% of patients, whereas autoimmune etiologies were identified in only 12% [129]. LPDs are frequent culprits for MAS/HLH, especially lymphomas (Fig. 3, E-F) [127, 129, 130]. The most common lymphomas to cause MAS/HLH are DLBCL, Peripheral T-Cell Lymphoma (PTCL) and Extranodal NK/T-Cell Lymphoma (ENKTCL). Reports of lymphoma-associated HLH show a relatively even distribution of B and T cell malignancies [131]. MCD can cause HLH in up to 30% of patients, especially the TAFRO syndrome variant [132, 133]. Kikuchi's disease may rarely cause a severe hyperinflammatory syndrome resembling MAS/HLH [53].

Currently there are few biomarkers available to distinguish between the different triggers for HLH, although small studies suggest that high IL-18 levels may be more common in autoimmune diseases, especially AoSD [134]. Further studies from different regions are needed to evaluate whether IL-18 alone or in combination with other clinical factors can be used to reliably distinguish rheumatologic from other secondary causes of HLH. Further clinical challenges include the availability of the test, as well as the time it takes for the test to result, when prompt empiric treatment is crucial. Initial treatment with high dose steroids has a potential to mask lymphoma and decrease the viability of biopsy specimens, although in our experience it is still possible to diagnose early-stage lymphoma on biopsy, even while the patient has been on a time limited course of high dose corticosteroids [135].

### **Hyper eosinophilia: Distinguishing Rheumatologic from Lymphoproliferative Causes**

Contemporary classification systems separate etiologies of eosinophilia into primary (neoplastic) or secondary (reactive) forms. Although mild or transient eosinophilia can be seen in many rheumatologic diseases, persistent hyper eosinophilia, defined as absolute eosinophil count exceeding  $1.5 \times 10^9/L$  ( $1,500/\mu L$ ) on at least two occasions separated by one month, is not a common occurrence in the rheumatology clinic. There is scarce literature to describe the prevalence of eosinophilia in rheumatologic disease. In our experience, EGPA, IgG4-RD, and Eosinophilic Fasciitis (EF) are the rheumatologic conditions that most commonly present with hyper eosinophilia. CTDs, RA, and AoSD are more likely to present with transient, mild eosinophilia. It is unlikely for autoimmune conditions to present with hyper eosinophilia without other systemic signs. However, certain features including low grade fever, B symptoms, and eosinophil-mediated damage (i.e. involvement of the skin, lungs,

GI tract, heart, or thrombotic events) are shared amongst many causes of hyper eosinophilia [136]. LPDs may be particularly challenging to differentiate from rheumatologic disease.

HL (Fig. 3, C-D), PTCLs, Kimura Disease (Fig. 2, I-L), and the lymphocytic variant of hyper eosinophilic syndrome (L-HES) are LPDs that can frequently cause hyper eosinophilia while mimicking rheumatologic disease [136–138]. Supplemental Table 5 offers guidance in differentiating LPD from rheumatologic etiologies of hyper eosinophilia.

### **Rheumatologic Manifestations of Lymphoproliferative Disease**

In this next section we discuss prominent manifestations of LPDs that when present will frequently lead to referral to a rheumatologist. Tables 1, 3, and 4 describes overlapping clinical (such as skin lesions), laboratory (such as positive serologies), and histopathologic (such as vasculitis) features between LPDs and rheumatologic conditions, based on review of recent literature [84, 139–146]. Supplemental Table 6 describes the primary LPDs that are known to be associated with each rheumatologic disease [147–150].

#### **Arthritis**

Lymphomas and leukemias can rarely cause paraneoplastic inflammatory arthritides [151, 152]. The hands, knees, and ankles are the most common joints affected [153]. Presentations can mimic RA (polyarticular, symmetric), seronegative spondyloarthropathy (oligoarticular, asymmetric) or remitting seronegative symmetric synovitis with pitting edema (RS3PE) (Table 4) [154, 155]. In our experience, patients with LPD-related arthritis often report an initial transient response to steroids potentially due to placebo effect, nonspecific pain relief, or natural fluctuation of symptoms. When pain recurs, clinicians may escalate steroid dosing with a similar response until the lack of consistent and objective improvement becomes apparent [156]. Elevated lactate dehydrogenase levels have been suggested as a distinguishing feature of paraneoplastic arthritis from hematologic malignancies compared with rheumatoid arthritis, but more research is needed to validate this [156]. Much more rarely, lymphomas can cause direct tumor invasion into the joint space which manifests as an inflammatory monoarticular arthritis mimicking septic arthritis [157, 158]. AoSD and aggressive T-cell lymphomas can both cause high fevers, diffuse LAD, diffuse rash, MAS, and abnormal large T-cell infiltrates on skin biopsy [159].

**Table 4** Selected rheumatologic manifestations of lymphoproliferative diseases

System	Feature	Most Common Rheumatologic Condition	Most Common LPD
Cardiac	Pericarditis	CTDs Seropositive RA	DLBCL
Pulm	Interstitial Lung Disease	CTDs Seropositive RA Sarcoidosis ANCA Vasculitis	MALT-L LYG MCD
Renal	Immune complex mediated glomerulonephritis	SLE	CLL PCD FL
MSK	Symmetric Inflammatory Polyarthritits	Any	DLBCL
MSK	Enthesitis, Dactylitis, Sacroiliitis	Seronegative spondyloarthropathies	ALL
Derm	Sclerotic skin thickening	Scleroderma	Scleromyxedema POEMS CTCL
Derm	Psoriasiform plaques	Seronegative spondyloarthropathies	CTCL
Derm	Panniculitis	Sarcoidosis Seronegative spondyloarthropathies SLE Behcet's	CTCL (SPTCL)
Neuro	Aseptic meningitis in the absence of systemic disease	Neurosarcoidosis CNS Vasculitis CNS IgG4-RD	PCNSL Bing-Neel Syndrome (WM) IVL LYG

In this table, we list the LPDs that most commonly mimic rheumatologic disease, organized by specific clinical features

*PCD* Plasma cell dyscrasia, such as multiple myeloma, WM, and AL amyloidosis, *POEMS* Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy, and Skin changes. This paraneoplastic syndrome is associated with PCDs and MCD, *ALL* Acute lymphoblastic lymphoma

## Cutaneous Lesions

Multiple cutaneous lesions and findings can mimic rheumatologic disease, often both clinically and histopathologically (Tables 3 and 4) [139, 160, 161]. Cutaneous lymphomas, particularly B cell lymphomas, may be annular, erythematous, and edematous and can mimic tumid lupus [162, 163].

Plaque stage cutaneous T cell lymphoma (CTCL) presents as psoriasiform plaques, which in the absence of biopsy can easily be mistaken for plaque psoriasis. It is important to note that even with biopsy, CTCL can be difficult to diagnose and that recent treatment (within 2 weeks) with oral

or topical steroids can mask the true findings. Median time from onset of symptoms to diagnosis of CTCL is 3–4 years [164].

SPTCL, particularly the alpha-beta subtype, can frequently have features both clinically and histopathologically resemble rheumatologic conditions [139]. Patients may present with fever, LAD, splenomegaly, and/or hepatomegaly along with cutaneous lesions that may resemble panniculitis (erythema nodosum, nodular panniculitis, lupus erythematosus profundus/panniculitis), systemic vasculitis, or pyoderma gangrenosum [139]. Differentiation may be difficult and require multiple biopsies, imaging, LN biopsies, and B cell and T cell receptor gene re-arrangement studies.

Angioinvasive or intravascular lymphomas can mimic medium vessel vasculitis or panniculitides, with tender subcutaneous nodules that progress to necrotic ulcers [165, 166].

## Central Nervous System Inflammation

Rheumatologic diseases that affect the central nervous system (CNS) include neurosarcoidosis, neuropsychiatric SLE, CNS Sjögren's syndrome, IgG4-RD, and CNS vasculitis [167]. A significant challenge in diagnosis has been establishing formal diagnostic criteria for each of these rheumatologic conditions that is not reliant primarily on obtaining CNS tissue. Furthermore, the pathogenesis of CNS involvement in each of these conditions is poorly understood.

Various LPDs can mimic rheumatologic disease in the CNS clinically, radiographically, and even histopathologically (Table 4). For late-stage lymphomas that metastasize to the CNS parenchyma or leptomeninges through hematogenous or local spread, peripheral LAD will be present, allowing for diagnostic LN biopsy to be performed. However, LPDs may also cause paraneoplastic CNS syndromes – HL can present with paraneoplastic granulomatous CNS vasculitis [168, 169].

Primary CNS lymphoma (PCNSL) can present as one or multiple mass lesions on MRI that are contrast enhancing, hypointense on T2 weighted sequencing, and cause diffusion restriction [170]. Metastases to the parenchyma and meninges can cause different imaging Patterns, including but not limited to parenchymal lesions or masses, leptomeningitis, and pachymeningitis. The most common subtype of PCNSL is DLBCL. In about 80–90% of patients with CNS lymphoma with leptomeningeal involvement, CSF studies will show pleocytosis and elevated protein [171, 172]. In atypical cases, CNS lymphoma can present as lymphomatosis cerebri, causing white matter lesions that may not enhance mimicking neurosarcoidosis or neurobehcet's when affecting the brainstem (Supplemental Fig. 3) [172]. CNS

lymphoma overlaps significantly in clinical presentation, neuroimaging, and even histopathology with neurosarcoidosis and CNS vasculitis [172–175]. For patients with history of DLBCL without known CNS involvement, relapse in the CNS occurs most frequently early after systemic therapy [49]. Relapse of CNS lymphoma should be ruled out before consideration of a new rheumatologic process.

Intravascular DLBCL (IVL) is an aggressive subtype of lymphoma that presents without LAD and can frequently involve the brain (Table 4). Diagnosis is challenging and often requires brain biopsy. Patients present with fever of unknown origin, constitutional symptoms, rash, and encephalopathy [176]. The neuroimaging MRI features include a T2-hyperintense lesion in the pons, nonspecific white matter lesions, mass-like brain lesions, multifocal areas of diffusion restriction appearing similar to infarcts, and meningeal enhancement [177, 178]. The areas of diffusion restriction may vanish or undergo the usual progression of changes seen in small vessel infarcts [179]. A hyperintense lesion in the pons is the most common neuroimaging finding and is also unique to intravascular lymphoma among LPDs affecting the CNS [178]. In the absence of a rash, there is potential for random skin biopsies to catch this subtype of lymphoma, especially when cherry angiomas are targeted [180–182]. Other than skin, bone marrow is the highest yield site for biopsy when considering this diagnosis [176, 183]. Overall, the neuroimaging and clinical presentation resembles primary CNS vasculitis and neurosarcoidosis [172].

Lymphomatoid granulomatosis (LYG) is an EBV associated LPD that can rarely present isolated to the brain, but more commonly presents with systemic involvement, including lungs, mediastinal LNs, CNS, liver, skin, and kidneys. Serum ACE is often elevated and biopsy results may be confused for sarcoidosis or vasculitis (Tables 1, and 4).

### Primary Immunodeficiencies Presenting as Lymphoproliferative Disorders with Rheumatologic Manifestations

Adult onset primary immunodeficiencies may not only cause recurrent infections, but also have autoimmune features and lead to development of lymphomas [184, 185]. These are important to distinguish, as initiation of immunosuppression may lead to significantly increased risk of infections. Although genetic diseases mostly arise in childhood, late onset disease is being increasingly recognized [186–188].

Common variable immunodeficiency (CVID) is the most common adult-onset primary immunodeficiency. Approximately 30% of CVID patients have autoimmune features resembling CTDs (immune thrombocytopenia, autoimmune hemolytic anemia, diffuse lymphadenopathy and/

or splenomegaly), seronegative spondyloarthropathies or Behcet's disease (psoriasis, enteropathy, uveitis, inflammatory arthritis, recurrent mucosal ulcers), and sarcoidosis (granulomatous interstitial lung disease, uveitis) [189, 190]. CVID should be considered in rheumatology patients who have lymphadenopathy and/or cytopenia in the absence of serologic positivity for rheumatologic disease, poor treatment response, and history of recurrent sinopulmonary or gastrointestinal infections [191, 192]. History of recurrent, severe, or atypical opportunistic infections prior to initiation of immunosuppression is an important clue. Serum immunoglobulin levels are low (IgG and either IgA or IgM) in CVID, and a poor antibody response to vaccinations is seen. Patients with CVID and autoimmune features should further receive genetic testing with next-generation sequencing panels or whole exome sequencing. Results of genetic testing may help to clarify diagnosis, understand prognosis, and select treatment [191, 193].

Autoimmune lymphoproliferative syndrome (ALPS) can closely mimic rheumatologic disease (CTDs such as SLE or Sjogren's), most commonly presenting with autoimmune cytopenias, splenomegaly and/or lymphadenopathy [194]. The presence of cytopenias is atypical for sarcoidosis, IgG4-RD, and AoSD (Supplemental Table 1). ALPS can cause MAS, and even cause organ specific autoimmune manifestations such as arthritis, rash, nephritis, uveitis, vasculitis, serositis, and interstitial lung disease [195]. In these patients the presence of CD4, CD8 double negative T cells (identifiable by flow cytometry) may be helpful, but serum immunoglobulin levels are frequently normal or high [195].

Several primary immunodeficiencies can prevent the ability to control EBV, resulting in lymphoproliferation and increased risk of EBV associated lymphomas [196]. These diseases can mimic CTDs such as SLE or Sjogren's, and cause pathologic findings mimicking sarcoidosis and vasculitis. Patients with TNFRSF13 mutations, especially the BAFF-variant, may develop SLE or seropositive RA [197]. Distinguishing features of other inborn errors of immunity causing lymphoproliferation and mimicking rheumatologic disease are detailed in Supplemental Table 7, but more research is needed in this area to determine appropriate indications for genetic testing.

### Conclusion

LPDs are a rare but important mimic of rheumatologic disease, and these two entities can co-occur. When the clinical presentation, laboratory results, pathologic findings, and initial steroid responsiveness of an LPD suggests rheumatologic disease, diagnosis is incredibly challenging. Clinicians should be aware that initial steroid responsiveness in

any manner, whether it be improvement in arthritis, rash, or lymphadenopathy, frequently occurs in LPDs.

In this review, we have described manifestations common to both LPDs and rheumatologic diseases and identified detailed features that should be considered as red flags to the rheumatologist. Collaboration across multiple subspecialists is frequently helpful to improve diagnostic accuracy. While imaging can be suggestive for a diagnosis, tissue diagnosis through LN or bone marrow biopsy in conjunction with flow cytometry and molecular testing remains essential. An understanding of the logistics of histopathology is important to prevent diagnostic delay or misdiagnosis by clinicians.

However, we also acknowledge that in some cases all red flags may be absent, or the patient history may be too complex to discern what is relevant or not from a diagnostic perspective. Further research is needed to find reliable biomarkers and other non-invasive tests to distinguish rheumatic disease from LPDs. Standardized evaluation protocols for assessing persistent fever with B symptoms, lymphadenopathy, splenomegaly, and cytopenias in patients with rheumatologic conditions can reduce diagnostic errors. The optimal timing for hematology referral remains unclear and deserves systematic investigation.

For practicing clinicians, maintaining suspicion for LPDs in atypical cases is crucial. Recognition of red flags followed by withholding steroids and obtaining appropriate tissue samples can prevent the diagnostic delays that compromise outcomes.

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**Supplementary Information** The online version contains supplementary material available at <https://doi.org/10.1007/s11882-025-01220-x>.

**Acknowledgements** The authors wish to thank Dr. Austin Rezig for his assistance with table formatting. We also appreciate feedback from Dr. Kirtan Patolia and Dr. Youssef Saklawi during conceptual discussions.

**Author Contributions** J.S. and B.K. conceptualized the review. J.S., B.K., J.L., and M.N. contributed to the literature search and reference selection. J.S., E.C., L.C., J.L., and M.N. drafted sections of the manuscript. E.C., L.C., D.C., E.L., and J.D. contributed to critical revisions and editing. J.S., E.C., L.C., D.C., and E.L. prepared the figures and summary tables. E.L. and J.D. provided supervision. All authors reviewed and approved the final manuscript.

**Data Availability** No datasets were generated or analysed during the current study.

## Declarations

**Competing interests** The authors declare no competing interests.

**Human and Animal Rights** This article does not contain any studies with human or animal subjects performed by any of the authors.

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