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CASE REPORT

PRIMARY SPLENIC DIFFUSE LARGE B-CELL LYMPHOMA WITH CD30 EXPRESSION: A RARE CASE REPORT

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ABSTRACT

Primary splenic diffuse large B-cell lymphoma represents a very uncommon manifestation within the spectrum of non-Hodgkin lymphomas, comprising approximately 1% of the total caseload. We report the case of an eighty-year-old male who presented with unintentional weight loss, fever, and night sweats. Laboratory studies revealed anemia, thrombocytopenia, and elevated inflammatory markers. Imaging demonstrated splenomegaly with a large hypodense lesion, while the mediastinal and hilar lymph nodes showed only mild uptake on positron emission tomography/computed tomography, which was interpreted as indicative of inflammation. Splenectomy revealed a necrotic mass measuring 13×12×10 cm that replaced most of the splenic parenchyma. Histology showed diffuse infiltration by large atypical lymphoid cells with immunoblastic morphology. Immunohistochemistry confirmed B-cell lineage (CD20, *PAX5*) with negativity for CD5, *BCL2*, CD10, and *c-MYC*. The Ki-67 index was markedly elevated (95%). Importantly, the tumor also exhibited aberrant CD30 expression, a finding reported in only a minority of cases of diffuse large B-cell lymphoma. While uncommon, CD30 positivity has been suggested in the literature to carry prognostic implications and may represent a biologically distinct subset. This case emphasizes the diagnostic and clinical significance of recognizing atypical immunophenotypic features in primary splenic diffuse large B-cell lymphoma.

Keywords: Diffuse large B-cell lymphoma, immunohistochemistry, non-Hodgkin lymphoma, splenic neoplasms

INTRODUCTION

Diffuse large B-cell lymphoma (DLBCL) constitutes the most frequently diagnosed subtype among non-Hodgkin lymphomas (NHLs) (1). Splenic involvement is observed in approximately 20% of NHL cases; however, primary splenic DLBCL (PS-DLBCL) is extremely rare, accounting for approximately 1% of all lymphomas (2). Iannitto and Tripodo (3) proposed that splenic lymphomas may present with diverse clinical features and can be classified into three categories: asymptomatic patients with isolated splenomegaly, splenomegaly associated with alterations in peripheral blood counts, and splenomegaly accompanied by constitutional symptoms and abdominal discomfort. PS-DLBCL is challenging to diagnose due to its non-specific clinical presentation, and optimal management generally involves splenectomy followed by immunochemotherapy. DLBCL usually expresses pan-B-cell markers; occasionally, CD30, a marker classically associated with Hodgkin's lymphoma, may also be expressed in a subset of DLBCL cases (4). Herein, we report a rare case of PS-DLBCL with aberrant CD30 expression, highlighting its clinical, radiological, and immunohistochemical features.

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CASE REPORT

An eighty-year-old male patient was admitted to the internal medicine clinic with unintentional weight loss of 23 kg over the previous two months, accompanied by night sweats and fever. Laboratory evaluation revealed thrombocytopenia (platelets: $108\times10^3/mm^3$; reference range: $150\text{-}400\times10^3/mm^3$) and anemia (hemoglobin: 8.3 g/dL; reference range for male: 14-17,5 g/dL), while the white blood cell (WBC) count was within normal limits (WBCs: $7.81\times10^3/mm^3$; reference range: $4\text{-}10\times10^3/mm^3$). Inflammatory markers were elevated, including C-reactive protein (CRP) (CRP: 122 mg/L; reference range: 0-5 mg/L), along with increased urea levels (urea: 95 mg/dL; reference range: 20-55 mg/dL).

Abdominal ultrasonography revealed splenomegaly (157×87 mm) with a solid lesion measuring 128×85 mm. Computed tomography (CT) demonstrated a hypodense lesion of approximately 10 cm extending inferiorly from the lower pole of the spleen. Bilateral hilar and mediastinal lymph nodes, the largest measuring 2 cm, were within physiological limits, as confirmed by ^{18}F-fluorodeoxyglucose (FDG) positron emission tomography/CT (PET/CT) The mild uptake observed in these nodes was interpreted primarily in favor of an inflammatory or granulomatous process (Figure 1A, 1B, 1C). In the hypodense mass extending approximately 10 cm inferiorly from the lower pole of the spleen, increased FDG uptake was observed, suggesting malignancy. Based on these findings, the splenic lesion was evaluated for malignancy, and splenectomy was performed.

On gross examination, the spleen measured 16×14×10 cm. On the cut surface, an irregularly bordered necrotic lesion measuring 13×12×10 cm was observed (Figures 2A, 2B). Microscopic examination revealed focal thickening of the splenic capsule. The splenic parenchyma was extensively infiltrated by atypical lymphoid cells with large nuclei, separated by fibrous septa containing broad areas of necrosis (Figure 3A). The infiltrating cells exhibited immunoblastic morphology, characterized by eosinophilic cytoplasm and atypical nuclei (Figure 3B). The surrounding parenchyma showed expansion of the red pulp and congestion.

Immunohistochemical analysis demonstrated that the atypical large cells expressed CD20 and *PAX5*, confirming a B-cell immunophenotype. CD5 negativity helped exclude mantle cell lymphoma and small lymphocytic lymphoma, while pancytokeratin negativity ruled out epithelial malignancies (Figure 4A). The diffuse growth pattern and the immunoprofile, particularly the absence of *BCL2* and CD10 expression, were not consistent with follicular lymphoma. Splenic marginal zone lymphoma (SMZL) was also considered in the differential diagnosis; however, the presence of large immunoblastic cells with a very high proliferative index (Ki-67: 95%), together with *BCL6*, MUM1, and *FOXP1* expression, was not consistent with SMZL, which typically demonstrates small to medium-sized marginal zone cells and an indolent immunophenotype. Aberrant CD30 expression was observed, whereas *c-MYC* expression was absent (Figure 4B). In the absence of peripheral or systemic lymph node involvement, the case was interpreted as primary splenic DLBCL, activated B-cell-like (ABC) type. An informed oral consent was obtained from the patient.

DISCUSSION

Primary splenic DLBCL is a distinctly uncommon clinicopathological entity, accounting for approximately 1% of all DLBCLs and less than 1% of NHLs (5). Although splenic involvement occurs in 20-40% of systemic lymphomas, true primary splenic disease is rare. Careful exclusion of systemic DLBCL and other entities, including T-cell/histiocyte-rich large B-cell lymphoma, SMZL, and peripheral T-cell lymphoma (PTCL), is therefore essential (4).

Typically, PS-DLBCL is diagnosed at a median age of sixty-four years (4). Our patient was eighty years old, representing the older end of the spectrum. Abdominal pain is the most frequently reported symptom (81%), followed by B symptoms (59%) (3, 4). Our patient presented with B symptoms, splenomegaly, anemia, and thrombocytopenia, broadening the clinical spectrum described in the literature. Furthermore, the hypodense splenic lesion detected on PET/CT correlated with underlying necrotic and fibrotic changes on pathological examination. According to the classification scheme of splenic lymphomas proposed by Iannitto and Tripodo (3), our patient can be categorized within the third group, characterized by splenomegaly associated with constitutional symptoms and abdominal discomfort.

Diffuse large B-cell lymphoma is characterized by large-cell morphology and a mature B-cell phenotype. It comprises two main subtypes: germinal center B-cell-like (GCB) and ABC. Although the 5th edition of the World Health Organization (WHO) Classification of Hematolymphoid Tumors states that the GCB/ABC classification has limited clinical impact, maintaining this distinction is recommended (6).

Histologically, PS-DLBCL is characterized by large atypical lymphoid cells with vesicular chromatin and prominent nucleoli (1). Immunohistochemical analysis typically demonstrates pan-B-cell markers such as CD20 and CD79a (7). In our case, the tumor cells expressed CD20 and *PAX5*, confirming B-cell lineage, and were negative for CD5, *BCL2*, and CD10, thereby excluding mantle cell lymphoma, small lymphocytic lymphoma, and follicular lymphoma. A more detailed evaluation of the differential diagnoses reveals that the most common clinical manifestations of mantle cell lymphoma are B symptoms or symptoms related to the involved lesion, although patients may also be asymptomatic at presentation. In addition, lymphocytosis may accompany the clinical findings. Mantle cell lymphoma typically demonstrates a CD5-positive B-cell phenotype characterized by expression of CD19, CD20, surface immunoglobulin M (IgM)/immunoglobulin D, and FMC-7, with light-chain restriction, reduced or absent CD23 expression, and strong cyclin D1 overexpression. In appropriate clinical settings, molecular workup may involve the determination of immunoglobulin heavy chain gene somatic

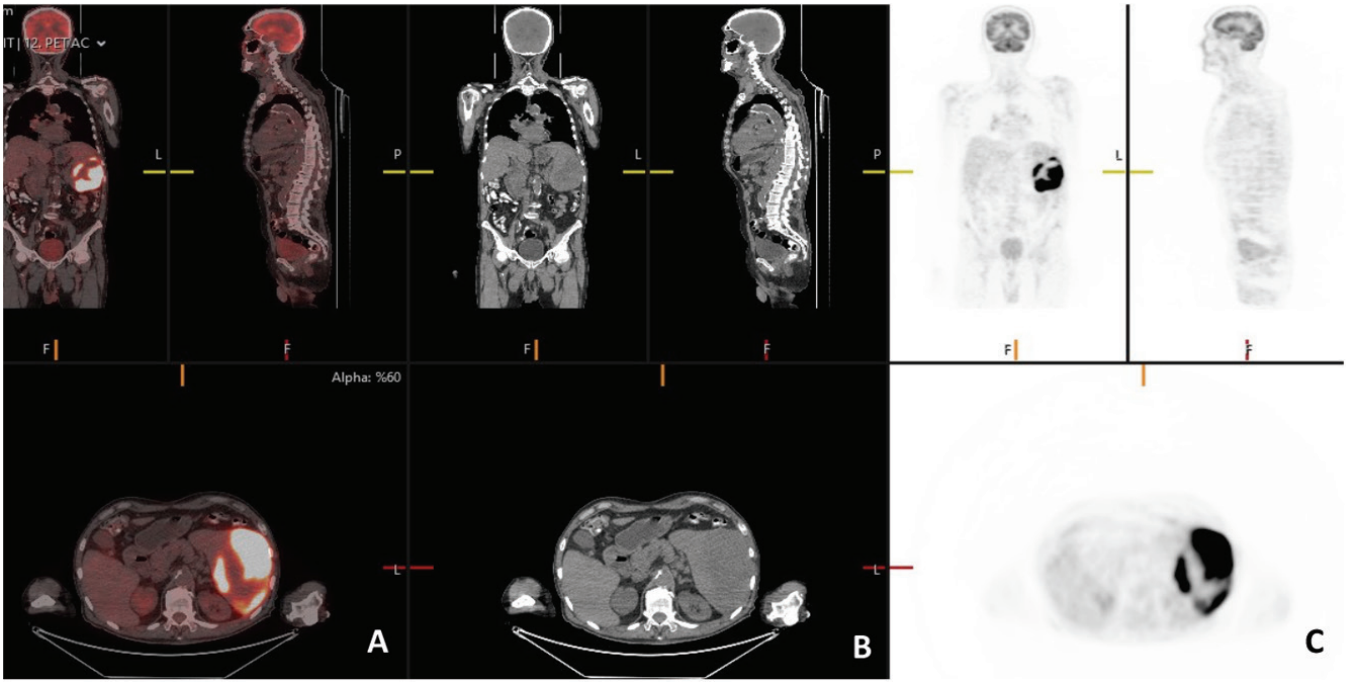

Figure 1: PET/CT (A), CT (B), and 18F-FDG PET/CT (C) scans show increased uptake localized to the enlarged spleen (SUV_{max} = 36.9), findings consistent with lymphoma.

PET/CT: Positron emission tomography/computed tomography, ^{18}F-FDG: ^{18}F-fluorodeoxyglucose, SUV_{max}: Maximum standardized uptake value

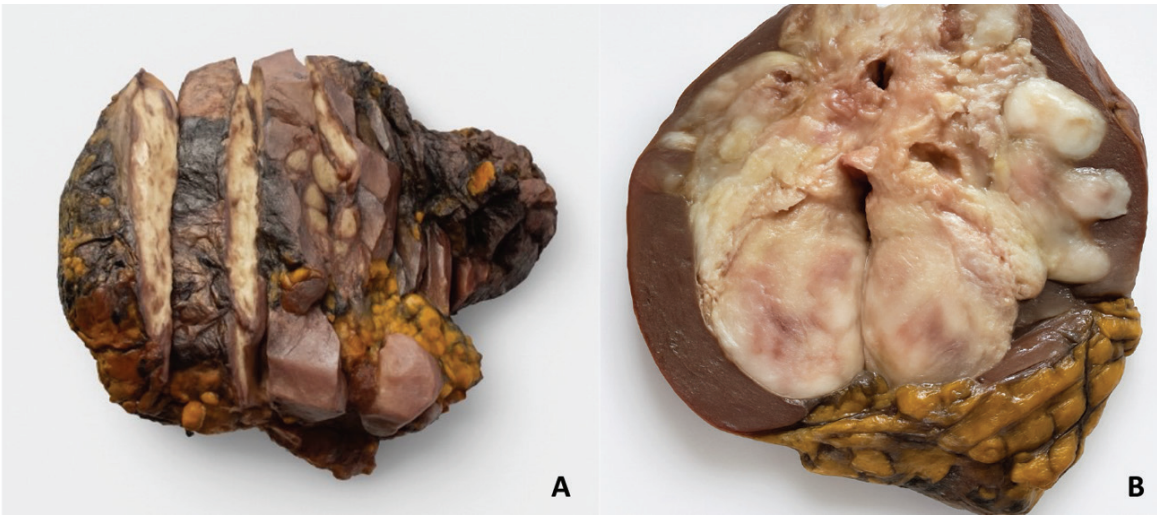

Figure 2: Gross appearance of the spleen. Increased weight and size with nodular areas on the outer surface of the capsule (A) and a solid mass with dirty-white nodular areas replacing the splenic parenchyma on the cut surface of the spleen (B) are shown.

mutation status and/or targeted next-generation sequencing for recurrent genomic alterations. Mutations involving *TP53*, *NOTCH1/2*, *SMARCA4*, *NSD2*, and *CCND1* have prognostic significance (8). Follicular lymphoma frequently presents with cervical or abdominal lymphadenopathy. While most patients are asymptomatic, symptomatic individuals may present with B symptoms and recurrent infections. Laboratory results are frequently unremarkable. Microscopic examination reveals incomplete or complete disruption of the lymph node architecture with multiple, comparably sized, nonpolarized neoplastic follicles and a thinned or missing mantle zone. Follicular lymphoma cells are positive for CD19, CD20, CD22, CD79, *PAX5*, and monotypic surface Ig (especially IgM). They also demonstrate germinal center markers such as CD10, *BCL6*, HGAL, *LMO2*, *STMN1*, GCET, and *MEF2B*. Commonly, cytogenetic analysis of follicular lymphoma cells identifies IGH-

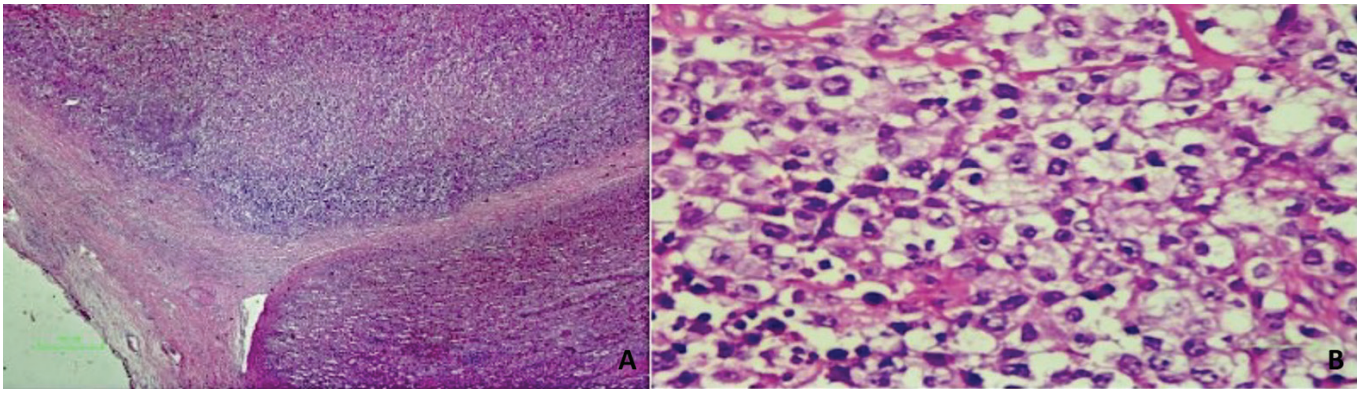

Figure 3: Microscopic examination. Low-power photomicrograph demonstrating nodular and diffuse infiltration of atypical lymphoid cells involving the splenic parenchyma and extending to the thickened capsule (H&E, ×40) (A). Microscopic view of diffuse large B-cell lymphoma demonstrating diffuse infiltration by large atypical lymphoid cells with immunoblastic morphology, vesicular chromatin, and prominent nucleoli (H&E, ×400) (B).

H&E: Hematoxylin and eosin

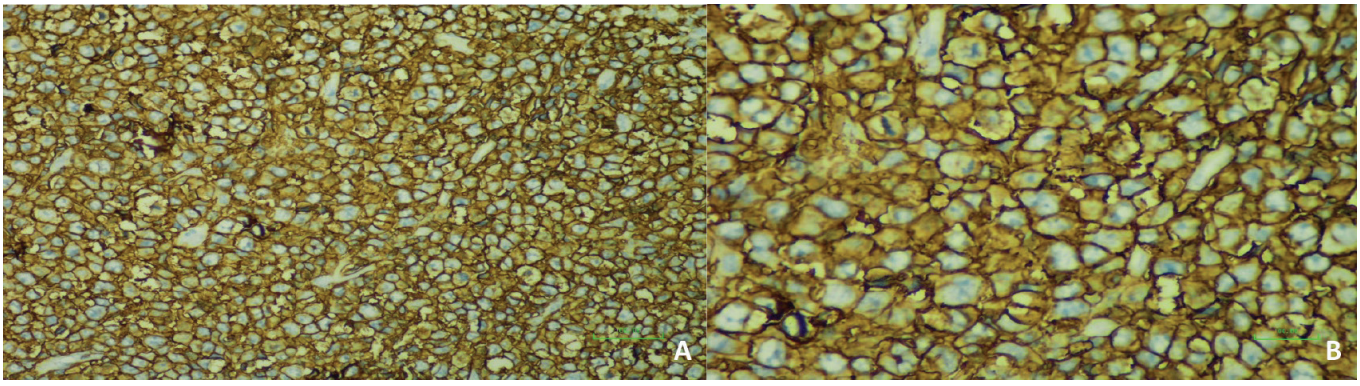

Figure 4: Immunohistochemical staining confirmed diffuse strong membranous CD20 expression in tumor cells (IHC, ×200) (A). Immunohistochemical staining confirmed strong membranous and cytoplasmic CD30 expression in tumor cells (IHC, ×400) (B).

IHC: Immunohistochemistry

BCL2 translocation, t(14;18) (q32;q21) (9). The clinical features of SMZL commonly include splenomegaly with bone marrow and peripheral blood involvement. Immunohistochemical examination demonstrates that SMZL cells express B-cell markers such as CD20 and *PAX5*, and are commonly positive for IgG and IgM and negative for germinal center markers. Genetic and molecular findings show that SMZL genetic abnormalities include alterations in apoptosis regulation, BCR and TNF signaling pathways, and nuclear factor kappa B (NF-κB) activation, involving genes such as *SYK*, *BTK*, *BIRC3*, *TRAF3*, *TRAF5*, *CD40*, and *LTB*. Differential diagnosis is guided by the examination of peripheral blood and bone marrow (10). In PTCL, patients present with constitutional symptoms. Microscopic examination reveals heterogeneous morphology and predominantly medium-sized or large cells with irregular nuclei and prominent nucleoli. Immunohistochemical analysis demonstrates that PTCL cells express pan-T-cell markers (CD3, CD2, CD5, CD7). Lack of *BCL2* expression, observed in 45-60% of cases, is considered a predictive marker of T-cell malignancies such as PTCL. Common genetic abnormalities include deletions of *CDKN2A* and *PTEN* (11).

In our case, aberrant expression of CD30 was observed, which is a feature reported in approximately 14% of DLBCL cases (12). While CD30 is classically associated with Hodgkin lymphoma and anaplastic large cell lymphoma (12, 13), its expression in DLBCL is less common and may have potential prognostic implications. In addition to its biological and prognostic implications, another important aspect of CD30 positivity is its role in the differential diagnosis between anaplastic lymphoma kinase (ALK)-positive large B-cell lymphoma and DLBCL. ALK-positive large B-cell lymphomas characteristically lack CD30 expression and are also frequently negative for CD20 (14).

Historically, splenectomy played a central role in both diagnosis and treatment of PS-DLBCL. With the introduction of rituximab, however, splenectomy is now primarily a diagnostic tool, while immunochemotherapy such as R-CHOP remains the standard of care (3, 15). Moreover, CD30 expression is not only relevant for differential diagnosis, but also for treatment plans. CD30 is a transmembrane glycoprotein exhibited by natural killer cells, dendritic cells, regulatory T-cells, and activated T-cells, and belongs to the tumor necrosis factor receptor superfamily (16). In light of its molecular structure, CD30 expression has shaped

new therapeutic strategies, such as the antibody-drug conjugate brentuximab vedotin (BV). In addition, studies have shown that CD30-positivity reflects important biological features, since high CD30 expression in tumor cells is associated with a characteristic immune landscape consisting of higher T-cell infiltration compared to CD30-low tumors. Additionally, within the tumor microenvironment, CD30 expression may influence the development of immunosuppressive or immunotolerant conditions (17). Based on these biological and pharmacological features, CD30-positive lymphomas have been associated with improved survival in some studies. Advances in CD30 targeted therapies consist of monoclonal antibody monotherapy, immunoconjugates (immunotoxins, radioimmunoconjugates, BV), bispecific antibodies, and CD30 chimeric antigen receptor T-cell therapy (18).

In our study, we used the Hans algorithm to classify the tumor as an ABC subtype, as it is the standard surrogate method recommended by the 5th edition of the WHO Classification of Hematolymphoid Tumors. To keep the diagnosis objective, we followed the common 30% cut-off for each marker. The subtyping was performed based on our immunohistochemistry results in a stepwise manner: first, the tumor cells were negative for CD10 (less than 30% expression), which ruled out the GCB subtype. Although *BCL6*-positivity was observed, the final classification was based on diffuse MUM-1 (*IRF4*) expression noted in the pathology report. Following the Hans decision tree, since the cells were CD10-negative and MUM-1-positive, the case was confirmed as a non-GCB/ABC phenotype (6). The ABC subtype is associated with a more adverse outcome following standard R-CHOP-based therapy compared with the GCB subtype, largely due to its constitutive activation of the NF-κB signaling pathway (17). Additionally, the presence of strong CD30 expression in the ABC background is a significant finding. It may define a unique biological group and, more importantly, it renders the tumor a potential target for antibody-drug conjugates like BV (19). By using the Hans criteria, we did not just label the subtype; we identified a phenotype that is crucial for deciding on modern treatments like polatuzumab vedotin (20).

CONCLUSION

Primary splenic DLBCL is an exceedingly rare entity, accounting for approximately 1% of all lymphomas and often posing diagnostic challenges. The present case highlights the diagnostic importance of thorough histopathological and immunophenotypic evaluation, particularly in an elderly patient with aberrant CD30 expression. It also contributes to the limited literature on this uncommon disease.

Ethics

Informed Consent: *An informed oral consent was obtained from the patient.*

Footnotes

Conflict of Interest: *The authors declared no conflict of interest.*

Author Contributions: *Surgical and Medical Practices: M.A.M., D.T.T., Y.E.A., F.Ü., F.Ö.P., Concept: B.Ö., M.A.M., F.Ö.P., Design: B.Ö., M.A.M., Data Collection or Processing: D.T.T., F.Ü., F.Ö.P., Analysis or Interpretation: D.T.T., F.Ö.P., Literature Search: B.Ö., H.B.K., Writing: B.Ö., H.B.K., M.A.M., F.Ö.P.*

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