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Cyclin D1-negative and SOX11-positive B-cell lymphoma with CD5 and CD10 coexpression and *MYC* rearrangement: A diagnostic challenge

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A 66-year-old male presented with a large left renal/perirenal mass (9.7 cm) and diffuse lymphadenopathies. A biopsy showed diffuse proliferation of monotonous small to intermediate lymphocytes with irregular nuclei and condensed chromatin (upper left, 20×; upper middle, 400×). The background epithelioid histiocytes and hyalinized vessels were noted. By immunohistochemistry, the lymphocytes were positive for CD20, CD5 (upper right, 200×), CD10 (middle left, 200×), SOX11 (middle central, 200×), and BCL2, but were negative for cyclin D1 (middle right, 200×), cyclin D3, CD23, and BCL6. MYC was expressed by 10%–20% of the cells with heterogeneous intensities (lower left, 200×), and the Ki67 proliferation index was about 30% (lower middle, 200×). Cytogenetic studies revealed *MYC* rearrangement in 97% of cells by using *MYC* break-apart probes (lower right, 5′ probe in red, 3′ probe in green) but with no evidence of *IGH::MYC*, *IGK::MYC*, or *IGL::MYC* fusions by further evaluation by fluorescence in situ hybridization (FISH). Assessment for rearrangements by using *CCND1*, *CCND2*, *CCND3*, *BCL2*, *BCL6*, and *IGH* break-apart probes were all negative.

Based on the morphological and immunophenotypical features, this most likely represents a cyclin D1-negative mantle cell lymphoma with *MYC* rearrangement. SOX11 is a highly specific marker for mantle cell lymphomas, which is consistently negative in other mature B-cell lymphomas other than Burkitt lymphoma [1]. However, we were not able to demonstrate the presence of *CCND1/2/3* rearrangements by break-apart FISH probes. It has been reported that cryptic insertions of immunoglobulin (IG) light chain enhancers are associated with cyclin D2/3 overexpression in cyclin D1-negative mantle cell lymphomas. Although cyclin D3 was not overexpressed in our case, cyclin D2 immunohistochemistry was not available and its overexpression cannot be excluded. In addition, a minor subset of mantle cell lymphomas lacks *CCND1/2/3* rearrangements but instead shows cyclin E upregulation [2]. *MYC* rearrangement can be seen in about 1.5% of mantle cell lymphomas, which is associated with blastoid morphology, aggressive disease course, and CD10 expression (seen in our case) [3]. To date, all the reported *MYC*-rearranged mantle cell lymphoma cases are positive for cyclin D1 by immunohistochemistry. In our case, the lack of cyclin D1/3 expression and *CCND1/2/3* rearrangements by break-apart FISH probes, in conjunction with the aberrant CD10 expression, confers diagnostic challenge. In addition, *MYC* showed rearrangement with a non-IG partner gene in this case. The translocation partners in *MYC*-rearranged mantle cell lymphomas are not well characterized. It remains to be explored whether the aggressive features associated with *MYC*-rearranged mantle cell lymphomas are preferentially seen in patients with *IG::MYC* (Figure 1).

AUTHOR CONTRIBUTIONS

Mariko Yabe designed the study and analyzed the data. Mingfei Yan analyzed the data and drafted the manuscript. Yanming Zhang performed cytogenetic study. Ahmet Dogan supervised the study. All the authors revised the manuscript.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

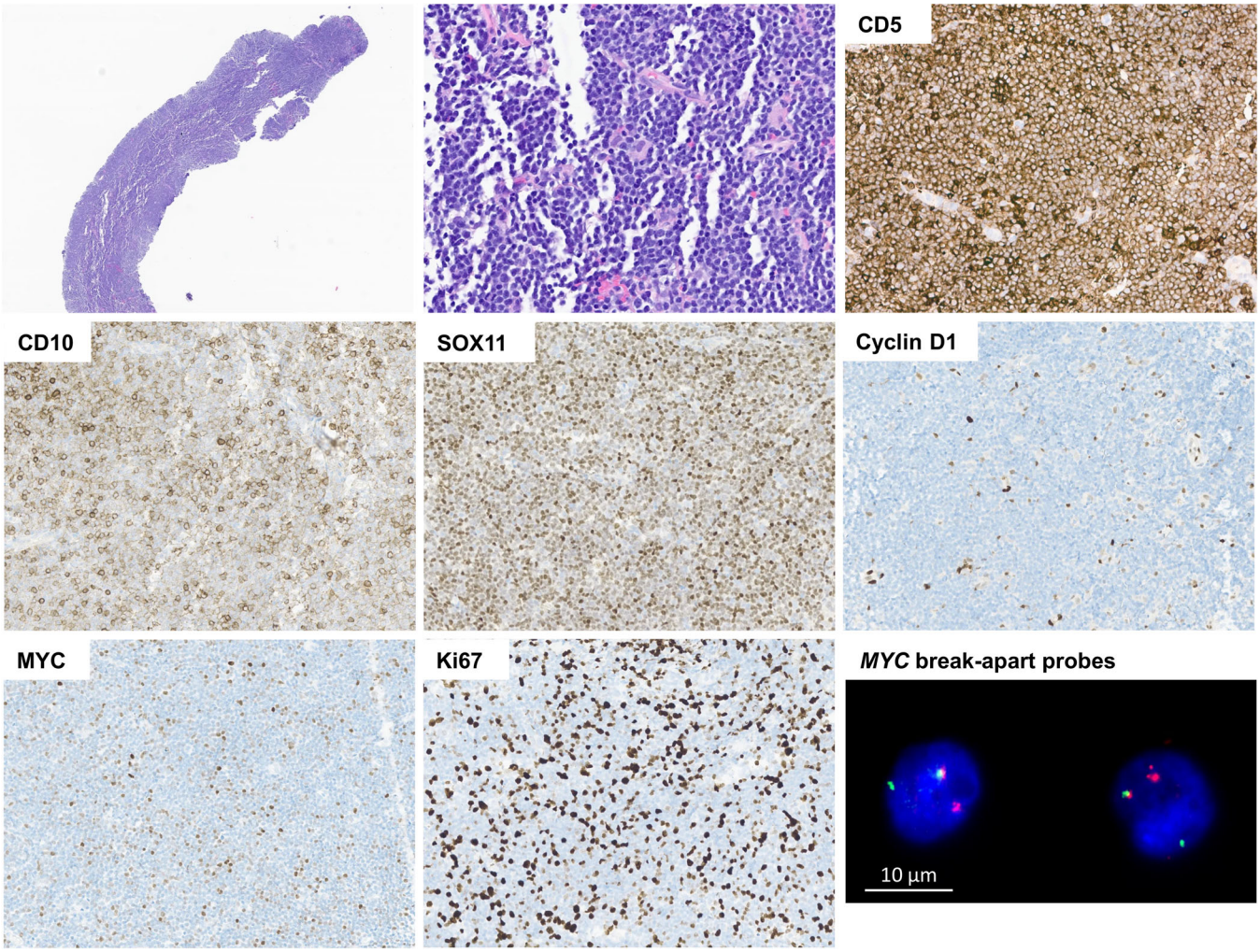

FIGURE 1 Upper left: Hematoxylin and eosin (H&E)-stained image of the core needle biopsy of the perirenal mass (magnification: 20×). Upper middle: H&E-stained image of the perirenal mass at higher magnification (400×). Upper right: The lymphoma cells were diffusely positive for CD5 (200×). Middle left: The lymphoma cells were diffusely positive for CD10 (200×). Middle central: The lymphoma cells were diffusely positive for SOX11 (200×). Middle right: The lymphoma cells were negative for cyclin D1 with only background stromal cells showing cyclin D1 positivity (200×). Lower left: The lymphoma cells were partially positive for MYC in 10%–20% of cells with heterogenous intensities (200×). Lower middle: The Ki67 proliferation index was about 30% in the lymphoma cells (200×). Lower right: Fluorescence in situ hybridization (FISH) image showing separation 5′ probe (red signal) and 3′ probe (green signal) in lymphoma cells by using *MYC* break-apart probes, consistent with *MYC* rearrangement.

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DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICS STATEMENT

This study is approved by our institutional review board.

PATIENT CONSENT STATEMENT

The authors have confirmed patient consent statement is not needed for this submission.

CLINICAL TRIAL REGISTRATION

The authors have confirmed clinical trial registration is not needed for this submission.

PERMISSION TO REPRODUCE MATERIAL FROM OTHER SOURCES

N/a.

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