66-year-old man presented with a white blood cell count of 62.6 \times 10^9/L. Peripheral blood (PB) smear showed 80% small- to medium-sized atypical lymphoid cells with irregular to cleaved (panel A; original magnification \times 500, Wright-Giemsa stain) or binucleated nuclear contours, and slightly condensed chromatin (panel B; original magnification \times 1000, Wright-Giemsa stain). Bone marrow (BM) aspirate (panel C; original magnification \times 500, Wright-Giemsa stain) and biopsy (panel D; original magnification \times 400, hematoxylin and eosin stain) showed \sim 70\% atypical lymphoid cells with similar features to those of PB. These lymphoid cells were positive for CD20 (panel E; original magnification \times 200) and CD5 (panel F; original magnification \times 100), negative for CD3 (panel G; original magnification \times 100) and cyclin D1 (not shown), but showed strong nuclear immunoreactivity for SOX11 (panel H; original magnification \times 400) and \sim 70\% proliferation index by Ki-67 (panel I; original magnification \times 200). Flow cytometry of PB and BM showed \lambda-restricted monotypic B cells, positive for FMC-7 but negative for CD23. Conventional karyotyping showed 46,XY[20], and fluorescence in situ hybridization analysis of BM showed no cyclin D1 or MYC rearrangements. A diagnosis of cyclin D1-negative blastoid mantle cell lymphoma (BMCL) was established.

This is an unusual BMCL with the following 3 features: initial leukemic presentation; markedly irregular to binucleated nuclear contours, which can be confused with other lymphomas such as follicular lymphoma or adult T-cell leukemia/lymphoma; and lack of cyclin D1 at both the protein and genetic level. Next-generation sequencing revealed KMT2D and TP53 mutations but no rearrangement of cyclin D2 or D3, respectively. Cyclin D1-negative MCL is difficult to recognize, but strong SOX11 nuclear immunoreactivity is helpful in this regard.

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Cyclin D1–negative blastoid mantle cell lymphoma exhibiting cleaved to bilobated cytomorphology

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