previous reports of thyroid PTCL-NOS, some had an aggressive course with the patient succumbing within 5−13 months despite chemotherapy and radiotherapy.[3] However, others reported spontaneous regression with subsequent disease-free survival of up to 97 months.[8] In this case, the decision was made for observation only due to the low disease burden.

In conclusion, this is a rare case of thyroid PTCL-NOS arising in a Caucasian male. Histologically, the features were like that of extranodal marginal zone lymphoma. Immunohistochemistry, flow cytometry and molecular studies were essential to clinching the diagnosis. It is yet to be elucidated if thyroid T-cell lymphoma is a distinct entity amongst the PTCLs-NOS. The natural history of reported thyroid T-cell lymphoma is variable and limited data are available on prognosis and treatment.

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## Primary splenic low-grade follicular lymphoma presenting with leukaemia and large cell transformation in the marrow

Sir,

Lymphomas in the spleen frequently represent secondary involvement by systemic lymphomas, while primary splenic lymphomas other than splenic marginal zone lymphoma (SMZL) and hairy cell leukaemia are uncommon and are poorly defined in the literature.[1] Follicular lymphoma (FL) is essentially a nodal disease and rarely occurs primarily in the spleen. Low-grade FL is an indolent disease but may transform to high-grade B-cell lymphoma [such as diffuse large B-cell lymphoma (DLBCL) and B-lymphoblastic lymphoma] during the disease course with a 10-year risk at around 28%.[2,3] Some of the transformed tumours may carry *MYC* and *BCL2* or *BCL6* rearrangement, so-called double-hit lymphoma.[4−7] Here we report an unusual case of primary splenic low-grade FL presenting synchronously as leukaemia and transformed large B-cell lymphoma (a double-hit lymphoma) in the marrow.

A 68-year-old male presented with fever for one month in July 2015. Haemogram showed leukocytosis (WBC at $31.4 \times 10^9$/L with 48.0% blasts and 25.0% lymphocytes) and normal levels of haemoglobin and platelets. Microscopic examination revealed large atypical lymphocytes with a fine chromatin pattern and prominent nucleoli (Fig. 1A). There was no peripheral lymphadenopathy. Abdominal ultrasound showed massive splenomegaly. Bone marrow aspirate showed numerous large atypical lymphocytes with similar morphology to that in the peripheral blood. Flow cytometric immunophenotyping of the marrow aspirate showed a mature B-cell phenotype with neoplastic cells expressing CD19 and CD20, and monotypic kappa light chain expression but not CD3, CD5, CD10, CD11c, CD23, CD43, CD103, or lambda light chain. Bone marrow trephine biopsy showed a diffuse infiltration of large lymphoid cells with frequent mitoses (Fig. 1B,C). The neoplastic cells expressed CD10, CD20 (Fig. 1D), bcl-2 (Fig. 1E), bcl-6, MUM1, and myc (Fig. 1F) with a high labelling index (90%) by Ki-67 immunostaining. Cyclin D1 and TdT were both negative. The marrow specimen was diagnosed as large B-cell lymphoma of germinal centre B-cell phenotype, and also a double-expressor lymphoma (positive for both bcl-2 and myc). Furthermore, bone marrow karyotyping revealed 48,XY,der(2q),der(3q),-der(6q),+7,+8,t(8;14)(q24;q32),der(13q),der(18q)[cp19]/46,XY[1], indicating translocation involving *IGH* and *MYC* genes.

As the spleen was the only involved organ by imaging studies, laparoscopic splenectomy was performed 10 days after marrow study, both for therapeutic and diagnostic purposes. There were no enlarged peri-hilar or abdominal nodes. The spleen measured 13 × 8.5 × 6.5 cm and weighed 498 g. A wedge of fresh splenic tissue was submitted for flow cytometric study and showed a similar immunophenotype as that of the marrow aspirate. Grossly, the spleen was beefy (Fig. 2A); but on closer inspection, the splenic parenchyma was replaced by numerous miliary whitish nodules of a slight variation in size (Fig. 2B). Histopathology showed a nodular/follicular pattern of infiltration centred in the white pulps

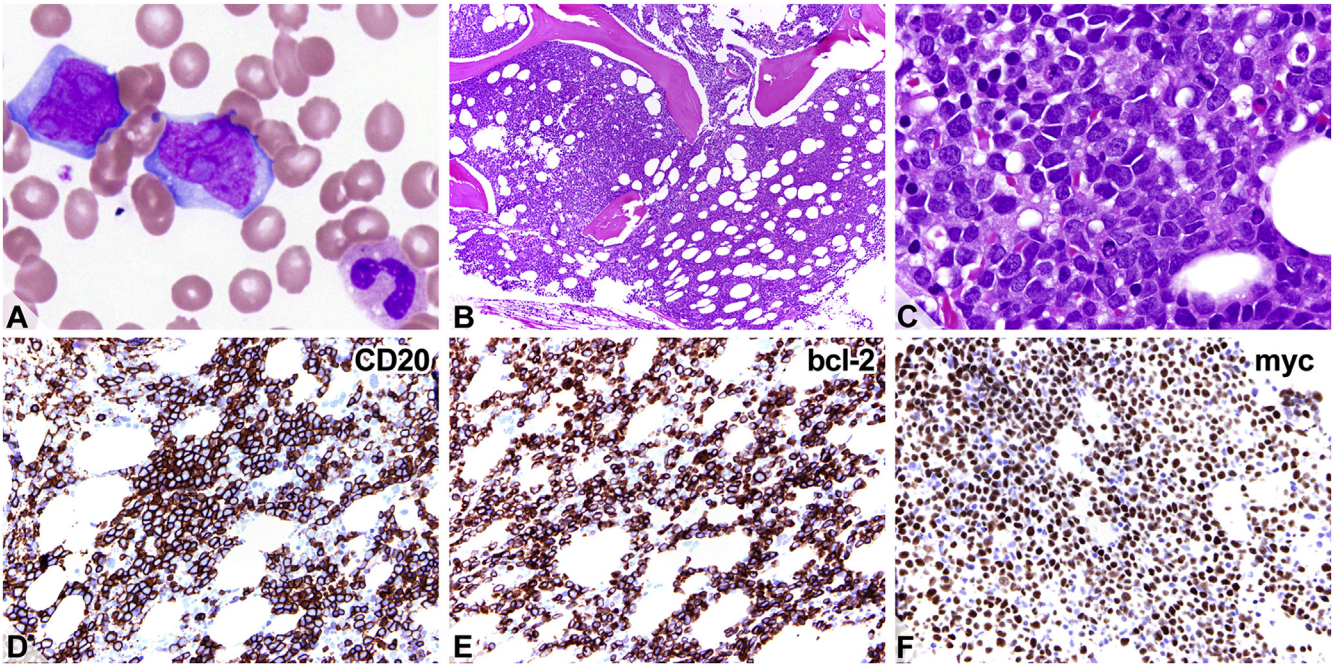


**Fig. 1** (A) Peripheral blood smear shows large atypical lymphocytes with prominent nucleoli. (B,C) Bone marrow trephine biopsy shows a diffuse infiltration of large lymphoid cells with frequent mitoses. The neoplastic cells express CD10, (D) CD20, (E) bcl-2, bcl-6, MUM1, and (F) myc.

(Fig. 2C). Under high power examination, these nodules were mainly composed of small to medium-sized centrocytes with occasional centroblasts (Fig. 2D). The neoplastic germinal centre cells expressed CD10, CD20, bcl-2, bcl-6, and kappa light chain but not CD3, CD5, cyclin D1, or lambda light chain. The Ki-67 labelling index was low (<10%). It was diagnosed as low-grade or grade 2 FL.

To evaluate the clonal relationship of the marrow and splenic tumours, B cell receptor gene rearrangement study was performed using the BIOMED-2 protocols. Both the splenic and marrow specimens were clonal with the tube A reaction of *IGK* gene showing the same band pattern (Fig. 3), indicating the same clonal origin of these two tumours. Interphase fluorescence *in situ* hybridisation (FISH) was performed as previously described.[8] The splenic FL showed rearranged *IGH* (Fig. 2E) and *BCL2* (Fig. 2F), but not *BCL6* or *MYC* with dual colour break-apart rearrangement probes (Vysis/Abbott Laboratories, UK). Reciprocal translocation of *IGH/BCL2* genes was confirmed by dual colour, dual fusion rearrangement probes (Vysis/Abbott Laboratories). These genetic changes were consistent with a low-grade FL. The marrow specimen showed rearranged *BCL2* locus and *IGH/BCL2* reciprocal

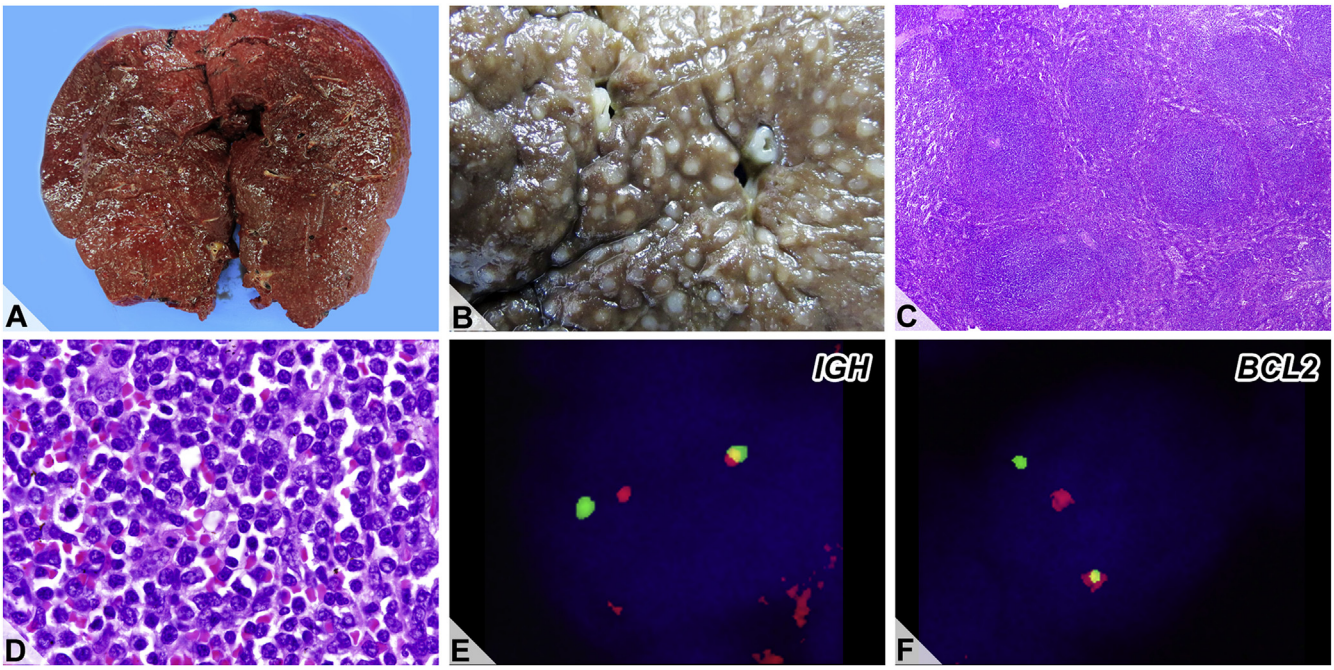


**Fig. 2** (A) Grossly, the spleen is beefy; (B) on closer inspection, it is replaced by numerous miliary whitish nodules. (C) Histopathology shows a micro-nodular pattern of infiltration in the white pulp, (D) comprising mainly of centrocytes. (E) FISH study shows rearranged *IGH* and (F) *BCL-2*, but not *MYC*.