



B Lymphoblastic Lymphoma/Leukemia with MYC Rearrangement

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Clinical Image

A 57-year old male with a history of type 2 diabetes mellitus presents with 6 months of fatigue, loss of weight and left neck lymphadenopathy. No immunodeficiency virus infection is noted. PET-CT shows left pharyngeal tonsil and cervical adenopathy, but no abnormal uptake in the chest and abdomen. Needle core biopsy of the left neck lymph node shows that the lymphoid tissue was diffusely infiltrated by medium-sized mononuclear cells with round to angulated nuclei, fine chromatin and small amount of cytoplasm (Figure 1A). Immunohistochemical staining showed that these atypical cells are positive for CD20 (subset) (Figure 1B), PAX5 (not shown), CD10 (strong) (Figure 1C), MYC (Figure 1D), and Terminal Deoxynucleotidyl Transferase (TDT, moderate) (Figure 1E), and are negative for BCL6, BCL2, cyclinD1, CD99, and CD34 (not shown). The proliferation index (Ki-67+) is 100% (Figure 1F). CD3 highlights a few background small T-cells (Figure 1G). Limited flow cytometric analysis detected a large population of surface light chain negative, CD45(dim)+CD19+CD20(dim)+ population. In addition, peripheral blood or bone marrow was not involved. FISH studies detected MYC rearrangement, but lacked rearrangement of BCL2 or BCL6 (H and not shown). B lymphoblastic lymphoma/leukemia was diagnosed. B lymphoblastic lymphoma/leukemia with isolated MYC rearrangement is extremely rare. According to a recent case report may represent the third case presenting as lymphadenopathy without bone marrow and peripheral blood involvement [1]. The two previously reported patients were treated with R-CHOP and hyper-CVAD, respectively, with good responses. Our patient is treated with R-Hyper-CVAD (cycle 2) with good response, further substantiating the notion that a treatment approach for Burkitt lymphoma rather than B-ALL/LBL may be warranted in these patients.

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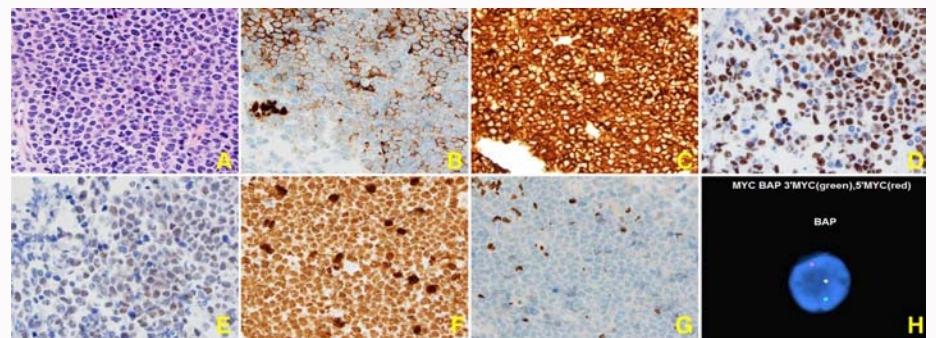


Figure 1: A) Fine chromatin and small amount of cytoplasm, B) Immunohistochemical staining showed that these atypical cells are positive for CD20(subset), C) PAX5(not shown), CD10(strong), D) MYC, E) terminal Deoxynucleotidyl Transferase (TDT, moderate), F) The proliferation index (Ki-67+) is 100%, G) CD3 highlights a few background small T-cells, H) FISH studies detected MYC rearrangement, but lacked rearrangement of BCL2 or BCL6 (and not shown).

References

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