DIAGNOSIS

Based on the histomorphologic and immunohistochemistry findings, a diagnosis of ALK positive large B cell lymphoma was proposed.

DISCUSSION

ALK-positive DLBCL is a rare subtype of non-Hodgkin's lymphoma (NHL) with distinct morphologic and immunohistochemical features. Delsol et al were the first to describe seven cases of diffuse large B cell lymphoma that expressed anaplastic lymphoma kinase in 1997.[1] This was recognized as a distinct entity in WHO classification of tumors of haematopoietic and lymphoid cells, in 2008.[2] It is known to occur in all age groups with male to female ratio of 3-5:1.[3] Lymph nodes are the most common site of involvement, but it is known to occur at the extranodal sites like tongue, nasopharynx, and stomach.[4]

This is a high-grade lymphoma that has a distinct morphologic appearance. The neoplastic cells are large, round, have centrally to eccentrically located nuclei, prominent central nucleoli, and scant to moderate amount of eosinophilic cytoplasm. Another important feature seen is a sinusoidal growth pattern. An extended immunohistochemical work up is essential to ascertain the lineage of these cells. These cells are usually negative for CD20, but focal positivity of Pax5 and CD79a helps to determine their B cell lineage. CD138 and single light chain cytoplasmic immunoglobulin A(IgA) are positive in the neoplastic cells.[5] CD30 is negative in the tumor cells along with T cell markers like CD3 and CD5. CD4 and CD43 are variably positive. The hallmark of these neoplasms is positivity for ALK. The staining pattern seen is either granular and cytoplasmic or membranous. Positivity for CD138, MUM1 and EMA suggests a plasmacytic differentiation. It is therefore thought that this neoplasm might be derived from post germinal B cell lymphocytes.[3] Also, in some cases cytokeratin (AE1/AE3) and EMA positivity may lead to erroneous interpretation of carcinoma. The negativity of biomarker CD30 can be used to differentiate it from ALK-positive Anaplastic large cell lymphoma, which is positive for ALK, CD30, and T-cell markers but negative for plasma cell marker (CD138) and immunoglobulin light chain. Positivity for ALK and CD138, also excludes Diffuse large B cell lymphoma, NOS. ALK positivity is unknown in Plasmablastic lymphoma, hence this helps us in differentiating these two entities.