Non-GCS (diffuse cytoplasmic, nuclear or nucleolar) pattern of ALK expression corresponds to translocation of ALK with different fusion partners like NPM1, SQSTM1, RANBP2 or EML4.[2]

ALK+ LBCL shows very aggressive clinical behaviour, a high relapse rate and a poor response to the standard treatment with cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) or CHOP-derived regimens. The overall survival (OS) is around 1 year in patients with advanced-stage diseases.[6,7]

Factors impacting prognosis include International prognostic index (IPI) score, stage and age at presentation. The underlying genetic abnormality and consequently the ALK staining pattern has also been reported to affect the prognosis.[8]

Currently, most patients are treated with CHOP/ CHOEP regimens and radiotherapy. Novel therapeutic approaches with anti-CD138 antibodies and ALK inhibitors may prove helpful in improving the prognosis of patients with this aggressive disease.

To conclude, knowledge of the disease entity with high index of suspicion supplemented with extensive immunohistochemical work up is required to make an appropriate diagnosis of this unique lymphoma.