# A Case of Diffuse Large B-cell Lymphoma with Pax-5 as the Only Positive B-cell Marker

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## INTRODUCTION

Pax-5 is a relatively new marker that encodes for BSAP (B-cell specific activator protein), a B-cell-specific transcription factor. Pax-5 is found in early stages of B-cell development and is required for B cell commitment. It plays an important role in B cell development, activation and differentiation. It is expressed in all stages of B cell development, but is down regulated at the plasma cell stage. In diffuse large B cell lymphoma (DLBCL), expression of Pax-5 parallels that of CD 20 and is lost in terminally differentiated tumors. We report an unusual case of pan-B cell marker negative (except for Pax-5 which was positive) DLBCL from pleural fluid. To our knowledge, this is the first reported case of DLBCL with neoplastic cells expressing Pax-5 as the only B cell marker.

## CASE HISTORY

This was a 79-year-old woman who presented with a one week history of worsening shortness of breath and pleural effusion. Peripheral blood showed iron deficiency anemia. CT scan revealed ill defined bulky soft tissue in region of the common iliac artery, mesentery, and iliac crest. Patient died 27 days after admission and on day 7 of chemotherapy.

## RESULTS

Pleural fluid contained large non-cohesive cells with immunoblastic features (Figs 1 and 2). Immunostains on cell block showed the neoplastic cells being negative for CD20 (Fig 3) and CD79a, and positive for Pax-5 (Fig 4). Additionally, tumor cells were positive for CD43, CD45, bcl-6, CD30 and CD10 (Fig 5-6); and negative for CD3, calretinin, pancytokeratin, S-100 protein, CD138, kappa and lambda immunoglobulin light chains. Flow cytometry was concordant with immunostains and showed the cells being negative for pan-B markers (CD19, CD20) and pan-T cell markers. In situ hybridization for EBV was strongly positive. As primary effusion lymphoma was a diagnostic consideration, cells were stained for human herpesvirus 8 (HHV8), which was absent. Patient was diagnosed with high-grade EBV-positive B-cell lymphoma with features of immunoblastic lymphoma.

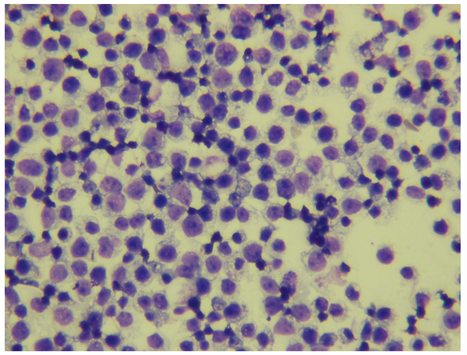

Fig. 1. Pleural fluid, Wright stain, 20X

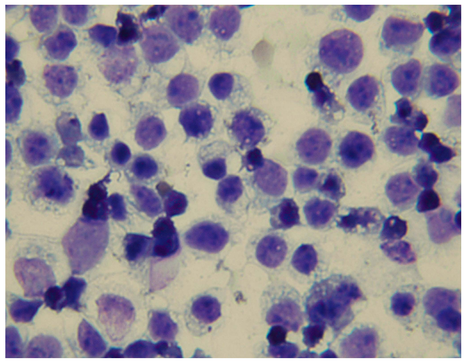

Fig. 2. Pleural fluid, Wright stain, 40X

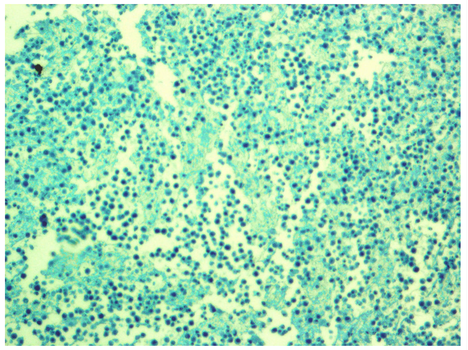

Fig. 3. Immunostain CD20, cell block, 10X

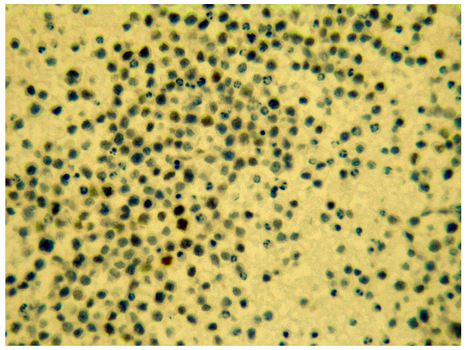

Fig. 4. Immunostain Pax-5, cell block, 10X

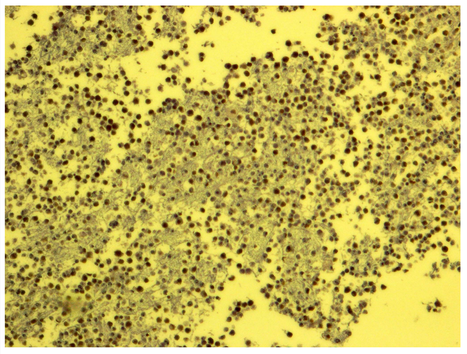

Fig. 5. Immunostain Bcl-6, cell block, 10X

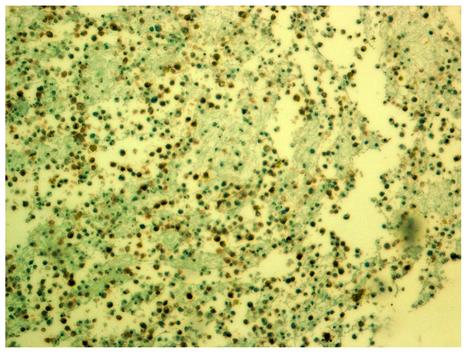

Fig. 6. Immunostain CD30, cell block, 10X

## DISCUSSION

CD20 and CD79a are the most widely used markers to establish B-cell lineage on paraffin sections. In cases of DLBCL with plasmablastic or immunoblastic morphology CD20 expression may be absent. However, in these cases cells typically express CD79a. Based on the location (pleural effusion), morphologic features, and negativity for multiple pan-B cell markers primary pleural effusion was a diagnostic possibility. Most cases of primary effusion lymphomas, however, are positive for HHV8. Activation markers (CD 30, CD38) and plasma cell-related markers (CD138) are also usually present. Another consideration was an unusual type of large B-cell lymphoma with immunoblastic morphology lacking B lineage markers. These tumors are referred to as DLBCL with expression of full length ALK and are consistently negative for CD 30 (the neoplastic cells in this case were positive for CD30). The aberrant expression of CD43 is part of the unusual presentation in this case.

## CONCLUSION

Among the B cell markers, the neoplastic cells were only positive for Pax-5. To our knowledge, this is the first reported case of DLBCL with the neoplastic cells expressing Pax-5 as the only B cell marker.

### References

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