Case of the Week

Hematopathology: Plasmablastic Lymphoma

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History

A 35 year old man presents to his physician with an enlarging neck mass. A core needle biopsy is performed.
Fig. 3. H&E stain, x400

Fig. 4. CD138 immunostain, x100
Fig. 5. Kappa, in-situ hybridization (ISH), x100

Fig. 6 Epstein Barr Encoding Region (EBER) ISH x100

**Diagnosis**

Plasmablastic Lymphoma
Discussion

Microscopic findings and immunohistochemistry

Sections show a diffuse sheet of large, pleomorphic cells with hyperchromatic nuclei, vesicular chromatin and prominent nucleoli. The neoplastic cells are reactive for CD138, EBV with kappa light chain restriction, but are negative for CD3, CD5, CD10, CD20, CD30, CD79a, desmin, EMA, ALK, AE1/AE3, CK5/6, lambda and HHV-8. Ki-67 demonstrated a high proliferative index (75%).

The differential diagnosis of this large cell lymphoid neoplasm includes diffuse large B cell lymphoma (DLBCL), plasmablastic lymphoma (PBL), primary effusion lymphoma, HHV8 associated large cell lymphoma and plasma cell myeloma. Negativity for HHV8 excluded primary effusion lymphoma and HHV8 associated large cell lymphoma and the absence of CD20 expression points against DLBCL. The association of EBV and a high Ki-67 index with this neoplasm favors PBL over plasma cell myeloma; even though immunoreactivity against CD138 with kappa restriction suggests a plasma cell neoplasm. Plasmablastic plasma cell myeloma must be considered in this case as it shares an almost identical immunophenotypical profile to PBL and has a high Ki-67 index, however, lacks the association with EBV. Furthermore, clinical correlation with skeletal surveys and serum protein electrophoresis is recommended when diagnosing PBL to assist in ruling out the possibility of plasmablastic plasma cell myeloma.

PBL is an aggressive non-Hodgkin lymphoma that is often associated with HIV infection. Originally described within the oral cavity, this cancer has since been described in many other body sites. Diagnosis of this entity can be challenging, since this lesion overlaps with plasma cell and lymphocytic neoplasms. The cells of origin are considered to be blastic post-germinal center B cells with a genomic profile that is similar to diffuse large B cell lymphoma (DLBCL). Approximately 50% of cases of PBL carry MYC gene rearrangements.

The prognosis of PBL is poor; without treatment, most patients die within 3-4 months. A standard chemotherapy protocol has yet to be established for PBL, however regimens such as EPOCH (etoposide, vincristine and doxorubicin with bolus cyclophosphamide and prednisone), usually prolongs life expectancy by several months. Therapies that target EBV may hold promise for better survival outcomes in the future.

References


