**Pediatric-Type Follicular Lymphoma: A Rare Entity with Excellent Prognosis**

**AUTHOR(S)**
Williams, Grant Capt

**PERFORMING ORGANIZATION NAME(S) AND ADDRESS(ES)**
59th Clinical Research Division
1100 Willford Hall Loop, Bldg 4430
JBSA-Lackland, TX 78236-9908
210-292-7141

**SPONSORING/MONITORING AGENCY NAME(S) AND ADDRESS(ES)**
59th Clinical Research Division
1100 Willford Hall Loop, Bldg 4430
JBSA-Lackland, TX 78236-9908
210-292-7141

**DISTRIBUTION/AVAILABILITY STATEMENT**
Approved for public release. Distribution is unlimited.

**SUPPLEMENTARY NOTES**
Texas Society of Pathologists Annual Meeting, Houston, Tx, Jan 20-21, 2018

**ABSTRACT**

**SUBJECT TERMS**

**SECURITY CLASSIFICATION OF:**
a. **REPORT**

**LIMITATION OF ABSTRACT**

**NUMBER OF PAGES**

18. **NUMBER OF PAGES**

19a. **NAME OF RESPONSIBLE PERSON**
Clarice Longoria

19b. **TELEPHONE NUMBER**
210-292-7141
A 13 year old female presented with a six month history of isolated supravcavicular lymphadenopathy. The patient had no constitutional symptoms and no significant past medical history. She had a non-diagnostic fine needle aspiration followed by an excisional biopsy. Part of the lymph node was sent for flow cytometry in RPMI and the rest of the tissue was fixed for histological, immunohistochemical, cytogenetic and FISH interpretation.

Histopathology

The lymph node was a 2.6 x 1.9 x 0.9 cm tan and pink tissue fragment with focal areas of hemorrhage. Histology showed an enlarged lymph node with architecture that was mostly effaced by a proliferation of large, expansive, and irregular follicles. The follicles had prominent admixed tingible body macrophages. Some areas showed marginal zone differentiation with pale appearing cells. The cells in the follicles were monotonous intermediate to large cells with finely clumped chromatin and small nucleoli. Some residual normal lymph node with reactive appearing follicles were present at the edges of the node.

Immunohistochemistry

Immunohistochemistry showed that the abnormal follicles were positive for CD20, CD10, BCL6, and dim CD43. The Ki-67 rate within the follicles is greater than 90% without polarization. The abnormal follicles were negative for BCL2, MUM1, and EBER in situ hybridization. CD21 and CD23 highlighted intact follicular dendritic cell meshwork in the areas of large B-cells. CD3 highlighted background T-cells.

Flow Cytometry, In Situ Hybridization, and Cytogenetic

Flow cytometry showed a CD10+ kappa immunoglobulin light chain restricted B-cell population comprising 52% of cells. There was no abnormal T-cell population identified. Fluorescent in situ hybridization showed negativity for MYC, BCL2, and BCL6 rearrangements.

The patient's cytogenetics revealed normal female karyotype: 46, XX.

Discussion

This constellation of findings is diagnostic and typical of pediatric-type follicular lymphoma, a rare variant of follicular lymphoma first recognized by the WHO in 2008. Follicular lymphoma is common in older adults but rare in pediatric and young adult patients. Pediatric follicular lymphoma comprises a only 6.5% of childhood lymphomas, has a median age range of 7.5-11.7, and has a male-female ratio of 4:1. This entity occurs most frequently in the tonsils and head and neck lymph nodes. The pathogenesis of follicular lymphoma in adults is based upon immunoglobulin gene rearrangements with BCL2, BCL6, and MYC. Pediatric follicular lymphoma is defined by a localized high grade appearing lymphoma that lacks these gene rearrangements. Other diagnoses to rule out are follicular hyperplasia and marginal zone lymphoma. Differentiation from follicular hyperplasia can be based off of total or extensive replacement of nodal structure, even distribution of nodules throughout the node as opposed to predominantly cortical distribution of follicles, crowding of nodules with little interposed lymphoid tissue, overall uniformity in size and shape of nodules, and paucity of reactive lymphoid cells in interfollicular areas. Ablent CD43 positivity also suggests neoplasia versus follicular hyperplasia. Differentiation from marginal zone lymphoma can be based of CD23 positivity and presence of follicular dendritic cells. Other immunohistochemical characteristics of pediatric follicular lymphoma are occasional CD10 positivity and high Ki-67 rates without polarization. Follicular lymphoma in pediatric patient's usually presents with grade 3 histology. When follicular lymphoma in young patients can be classified as pediatric follicular lymphoma based upon IHC findings and negative PCR of immunoglobulin gene rearrangement, as it was in 34 of 63 cases of follicular lymphoma in children in a 2009 study, the patient outcomes were better and there was a lower clinical stage at diagnosis.

Conclusion

Despite the high grade histology, the prognosis for pediatric follicular lymphoma is excellent and excisional biopsy is curative. Recognition of this rare entity is critical to guiding optimal clinical management and sparing patients from unnecessarily aggressive management. Had this patient's cytogenetic studies shown a rearrangement of BCL2, BCL6, or MYC than the diagnosis would have been follicular lymphoma, grade 3b. This diagnosis was managed surgically and the patient's disease has not recurred after 4 months of follow up.

References
