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FOLLICLE CENTER LYMPHOMA (FCL) OF THE LOWER FEMALE GENITAL TRACT (LFGT): A NOVEL VARIANT OF PRIMARY CUTANEOUS FOLLICLE CENTER LYMPHOMA (PCFCL)

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INTRODUCTION

- Follicle center lymphoma (FCL) of the lower female genital tract (LFGT) is a newly identified variant of primary cutaneous follicle center lymphoma (PCFCL).
- This rare lymphoma predominantly involves the uterine cervix and vagina, posing diagnostic difficulties due to its uncommon presentation and atypical features.
- The prognosis for FCL of the LFGT is generally favorable, with reports showing an 83% rate of durable complete remission and a 100% 5-year overall survival rate[1].
- Awareness of FCL is crucial to ensure accurate diagnosis and appropriate management.

AIM

- To illustrate the clinical, histopathological, and molecular features of FCL of the LFGT, emphasizing its rarity and distinct presentation.
- To discuss the difficulties in accurately diagnosing FCL of the LFGT due to its unusual location, histological resemblance to other conditions, and atypical immunohistochemical findings.
- To promote awareness about FCL of the LFGT, ensuring it is considered in differential diagnoses to prevent mismanagement and improve patient outcomes.

CASE DESCRIPTION

- A 49-year-old female with a Mirena intrauterine device experienced daily vaginal bleeding and postcoital bleeding 10 months post-placement.
- Initial imaging via pelvic ultrasound and CT scan revealed a 6 cm solid mass in the pelvis, near the vagina and lower uterine segment.
- A cervical biopsy initially suggested an atypical fibroblastic lesion resembling neurofibroma, with a proliferative index (Ki-67) over 20% and focal S-100 staining, indicating low-grade sarcoma.
- However, a repeat biopsy for a second opinion identified the lesion as large B-cell lymphoma with follicle center cell origin (*Image 1*). This was confirmed by immunohistochemical markers (CD10 and BCL6) and a high Ki-67 index (80%) (*Image 2)*. IGH/BCL2 FISH was negative.
- PET/CT imaging characterized the mass as a large, bulky soft tissue lesion centered at the cervix, indicative of aggressive lymphoma.
- Following a multidisciplinary tumor board discussion, the patient began an R-CHOP regimen (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) for 4 cycles, which resulted in significant improvement in vaginal bleeding after the first cycle.

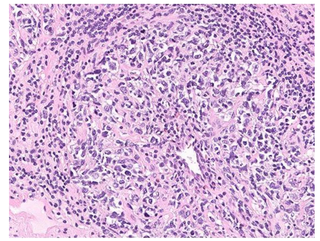


Image 1: Loosely arranged large lymphoid cells consistent with centrocytes and centroblasts cluster.
Source: The American Journal of Surgical Pathology[2]

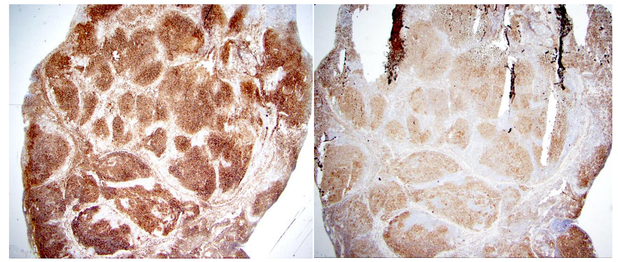


Image 2: Immunostain for BCL6 and CD10 highlights neoplastic lymphoma cells of germinal center origin within neoplastic follicles.
Source: PathologyOutline[2]

DISCUSSION

- FCL of the LFGT is a distinct, localized B-cell lymphoma similar to PCFCL in its histological, molecular, and clinical features.
- Histopathological and molecular diagnosis differentiate FCL from diffuse large B-cell lymphoma (DLBCL).
- Neoplastic cells in FCL are often CD20+ and BCL6+, BCL2 FISH negative, and exhibit a follicular pattern. Additionally, NGS has identified *TNFRSF14* as the most mutated gene, with significantly fewer mutations in *CREBBP* and *KMT2D.*
- The prognosis for FCL of the LFGT is generally favorable, with reports showing an 83% rate of durable complete remission and a 100% 5-year overall survival rate[1].

CONCLUSION

- Accurate diagnosis and a multidisciplinary approach are crucial for effectively managing rare lymphomas like follicle center lymphoma of the lower female genital tract, significantly impacting treatment outcomes and prognosis.

REFERENCES

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