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ALK-POSITIVE INFLAMMATORY MYOFIBROBLASTIC TUMOR WITH CONCOMITANT FOLLICULAR LYMPHOMA

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INTRODUCTION

- Inflammatory Myofibroblastic Tumors (IMT) are rare soft tissue tumors with approximately 150-200 new cases annually in the U.S.A[1].
- Bladder-localized IMTs are exceptionally rare, comprising less than 1% of all IMTs.
- Due to their rarity, standard treatment protocols are not well established.
- This report presents a case of an anaplastic lymphoma kinase-positive (ALK)-IMT compounded with follicular lymphoma.

AIM

- To highlight the rarity of Inflammatory Myofibroblastic Tumors, especially bladder-localized IMTs, and discuss the challenges in establishing standard treatment protocols due to limited cases.
- To emphasize the need for further research into the optimal treatment strategies for patients with multiple concurrent malignancies, like IMT and follicular lymphoma.

CASE DESCRIPTION

- A 38-year-old woman with a medical history of endometriosis and melanoma presented with hematuria and syncope.
- She experienced dysuria, urinary retention, and persistent hematuria despite treatment for a presumed urinary tract infection. Her hemoglobin was 8.3 g/dL.
- A computed tomography (CT) scan of the abdomen and pelvis revealed a bladder mass. Subsequent cystoscopy uncovered a 5 cm sessile "glistening" bladder tumor, which was resected.
- Pathological examination confirmed a spindle cell neoplasm consistent with ALK-IMT *(Image 1 & 2)*.
- CT scan of the chest revealed bilateral axillary lymphadenopathy.
- Excisional biopsy of the lymph nodes confirmed follicular lymphoma, Grade 1-2, with positive Kappa, CD 10, weak CD 19, CD 20, low CD 38, and CD 45 markers, and IGH-BCL2 fusion associated with t(14;18).
- The patient was managed with a 6-month cystoscopy while her follicular lymphoma, classified as Ann Arbor stage III, was clinically monitored.
- There was no evidence of IMT recurrence at 10 months follow-up.

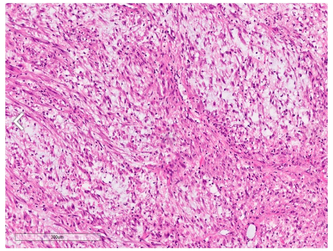


Image 1: Loosely arranged spindle cells growing in haphazardly arranged fascicles, interspersed by abundant chronic inflammatory cells (400x).
Adopted from Pathology Outlines[2]

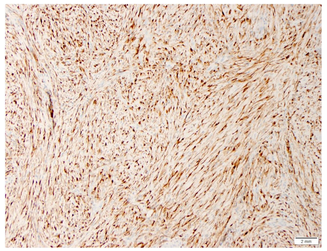


Image 2: Immunohistochemistry is positive for ALK1
Adopted from Pathology Outlines[2]

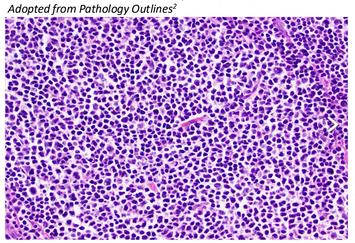


Image 3: H&E of lymph node showing neoplastic follicle is composed of numerous centrocytes and fewer centroblasts, compatible with grade 2 follicular lymphoma
Adopted from Pathology Outlines[2]

DISCUSSION

- Approximately 60-70% of IMTs exhibit ALK rearrangements. Surgical resection is the initial treatment for localized IMT, though recurrence can occur in up to 25% of cases[1].
- In phase I and II trials, crizotinib, an ALK inhibitor, has shown an overall response rate of 50%—86% for advanced and metastatic IMT. Other ALK inhibitors, such as brigatinib, lorlatinib, and alectinib, are also used in treatment-refractory IMT[3].
- The patient's follicular lymphoma was managed with surveillance due to a low-risk FL-IPI score and no criteria for immediate treatment.

CONCLUSION

- This case underscores the complexity of diagnosing and treating IMT with concurrent follicular lymphoma, emphasizing the need for a multidisciplinary approach and further research.

CONTACT INFORMATION

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