Reframing Breast Cancer Therapies for Metastatic Mammary-Like Vulvar Adenocarcinoma: A Case Report and Literature Synthesis

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Abstract

Mammary-like adenocarcinoma of the vulva (MLVA) is a rare subtype of vulvar adenocarcinoma that exhibits morphological and immunohistochemical features similar to breast tissue. Arising from anogenital mammary-like glands, this entity poses unique diagnostic and therapeutic challenges due to its extreme rarity and overlapping features with both breast and gynecologic malignancies. Currently, there are no standardized treatment guidelines for MLVA, and clinical management is often extrapolated from therapeutic strategies used in breast cancer, particularly for tumors with hormone receptor or HER2 expression.

Here we report a case of stage IV HER2-positive MLVA, which has been managed with a sequential systemic therapy regimens based on guidelines for metastatic HER2-positive breast cancer.

Introduction

- MLVA is a rare tumor with molecular similarities to breast carcinoma.
- Most tumors express estrogen and progesterone receptors, and approximately 10 to 15 percent demonstrate HER2 amplification, providing a biologic basis for adapting breast cancer–derived therapies.
- Genomic analyses show that MLVA aligns more closely with breast cancer than with other vulvar neoplasms.
- Current treatment decisions are largely extrapolated from established breast oncology regimens.

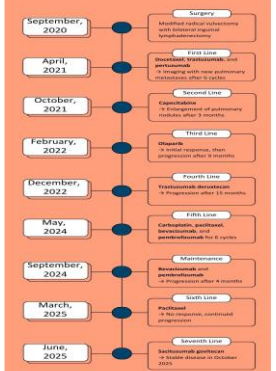
Case Presentation

Patient Profile
- Age: 54-year-old female.
- Presentation: Vulvar mass present for 3 years, recent growth causing discomfort as truck driver.

Initial Evaluation (2020)
- **Gynecologic Exam**: 5 × 3 cm firm left labia majora mass, 3 cm palpable inguinal lymph nodes.
- **CT pelvis**: 2.7 cm left vulvar mass, multiple enlarged bilateral inguinal lymph nodes.
- **PET-CT:** Hypermetabolic left labial neoplasm with metastatic retroperitoneal, left iliac, bilateral inguinal nodes.
- **Bilateral mammography**: Stable benign cystic changes.
- **Biopsy**: Left inguinal lymph node → metastatic adenocarcinoma.

Diagnosis & initial management
- **Stage IV** vulvar adenocarcinoma (pelvic lymph node involvement).
- **Surgical Intervention:** Modified radical vulvectomy + bilateral inguinal lymphadenectomy (Sept 2020).
- **Pathology:** Moderately differentiated adenocarcinoma from anogenital mammary-like glands, ER-/PR-, HER2-positive (IHC 3+).
- **Genetics**: BRCA2 pathogenic variant.
- Completed adjuvant radiation with concurrent weekly cisplatin.
- Subsequently referred to Breast Oncology for consideration of HER2-directed therapy.

Treatment History



Discussion

- Breast cancer–based strategies can be applied in MLVA due to molecular similarities.
- Hormone receptor–positive MLVA may benefit from endocrine therapy.
- HER2-positive MLVA can respond to trastuzumab-containing regimens.
- TROP2 overexpression provides rationale for antibody-drug conjugates such as Sacituzumab Govitecan.
- Diagnostic accuracy remains essential; Immunohistochemistry confirmation: GATA3, mammaglobin, GCDFP15 is critical.
- Inclusion in basket trials or registries for HER2- and TROP2-directed therapies is warranted.

Conclusions

- MLVA is an extremely rare malignancy with no established standards of care.
- Its histologic resemblance to breast cancer suggests it may respond to breast cancer-based therapies
- Case illustrates feasibility of therapy selection based on HER2 and TROP2 expression.
- Treatment decisions should be guided by receptor status and molecular markers.
- TROP-2-targeted mechanism of Sacituzumab Govitecan may offer a rational therapeutic approach in rare tumors with overlapping molecular features.
- Early recognition, accurate histopathology, and molecular profiling are essential for optimizing outcomes.

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References

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