

Rare Presentation of Primary Mucinous Adenocarcinoma, Mammary Type, of the Vulva

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Background:

- Primary mucinous adenocarcinoma of the vulva is an extremely rare disease, comprising only 1% of all vulvar cancers, with most primary vulvar cancers diagnosed as squamous cell carcinoma (90%).
- Primary mucinous adenocarcinoma of the vulva tends to arise from normal glandular structures, embryonic remnants, and ectopic tissues.

Case Description:

- 57-year-old G3P2012, with a 3.0-cm cystic tumor of the left labium majus for months
- No pain, pruritis, or discoloration.

Interventions:

- Vulvar biopsy with a histologic diagnosis of moderately differentiated mucinous adenocarcinoma, mammary type
- Initial breast work-up: mammography and core biopsy of hypoechoic avascular breast mass, with negative findings confirming the vulva as the primary site
- Partial radical hemivulvectomy and sentinel node biopsy
- Postoperative Tamoxifen therapy

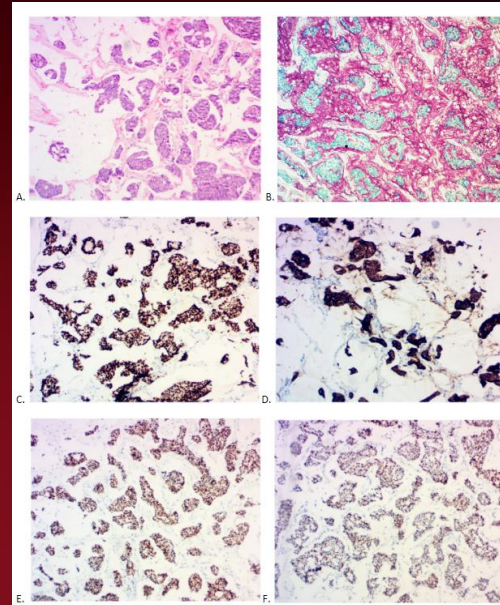


Figure 1: Primary Mucinous Adenocarcinoma, Mammary Type, of the Vulva: (A) Tumor cells in nests floating in pools of extracellular mucin (B) Extracellular mucin is highlighted on PAS-D histochemical stain. (C) Cytokeratin 7 immunostain is strongly positive in all tumor cells. (D) GATA-3 immunostain is strongly positive in all tumor cells, marker of mammary lineage. (E) Estrogen receptor immunostain is strongly positive. (F) Progesterone receptor immunostain is strongly positive.

Immunohistochemistry:

- Mucicarmine and PAS-D highlighted the abundant mucin present in the tumor.
- Tumor cells were diffusely positive for Cytokeratin-7 (CK7), GATA3, estrogen receptor (ER), and progesterone receptor (PR).
- The tumor was negative for HER2, CA125, Calretinin, and PAX8, excluding ovarian cancer origin, and negatively for CDX2 and CK20, excluding adenocarcinoma of the gastrointestinal tract.
- TTF-1 was also negative, thereby excluding lung origin.
- These resulting stains showed features which raised the possibility of metastatic breast cancer vs. an adenocarcinoma of mammary type, primary to the vulvar region.

Discussion:

- This patient's vulvar cancer likely originated from ectopic breast tissue, given the morphologic features and immunohistochemical profile.
- There has only been one reported case of a primary mucinous adenocarcinoma of the vulva arising from ectopic breast tissue, making this the second reported case.
- Due to the rare nature of primary mucinous adenocarcinoma of the vulva, mammary type, standardized postsurgical treatment and outcomes are unknown.
- Therefore, tumor tissue has been sent to Caris for genomic testing and will hopefully elucidate a targetable genomic profile for this exceptionally rare tumor.