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“Vulvar Paget’s disease: A single institution case-series in Louisiana “

Objective: Extramammary Paget’s Disease (EMPD) is a rare gynecological adenocarcinoma with a poorly understood pathophysiology affecting older women. Due to its rarity and complexity, no randomized control studies have been performed to establish standardized treatment or guidelines. This retrospective case-series aims to identify treatment protocols used for vulvar cancer patients presenting with EMPD at our institution and describe disease-free intervals and recurrence patterns to better understand the clinical characteristics of this rare and refractory disease and its management outcomes.

Methods: Patients diagnosed with vulvar EMPD from January 2015– June 2025 were identified using an electronic medical record database. Eleven patients were identified. Data abstraction of eleven medical records was completed using an IRB-approved gynecological oncology database within our institution. Data pertaining to patient demographics, disease diagnosis and characteristics, treatment protocols, and recurrence rates were collected.

Results: The mean age at diagnosis was 70.91 years old. Of the eleven patients who were identified, ten (91%) were Caucasian and one (9%) was Black/African American. All (100%) were non-Hispanic. Ten (91%) patients underwent surgery as their primary treatment, and one was prescribed imiquimod as a conservative first-line treatment due to co-morbidities and patient preference. Of those that underwent surgery, two (20%) had a partial vulvectomy, one (10%) had a radical vulvectomy, and seven (70%) had a vulvectomy (not otherwise stated). The patient who had a radical vulvectomy also underwent groin lymph node dissection. Margins were positive for four (40%), negative for three (30%) and unknown for three (30%). After their initial therapies, eight (73%) experienced a recurrence and all eight had surgery as their primary therapy. Additionally, five (45%) experienced only one recurrence, one (9%) experienced four total recurrences, one (9%) experienced six total recurrences, and one (9%) experienced seven recurrences. Nine (82%) had a disease-free interval of 0-3 years after surgery, including three patients who did not have biopsy-proven recurrence and are currently disease-free; 2 (25%) had a recurrence greater than three years after their surgery.

Conclusion: This retrospective case-series highlights the difficulty in defining a proper treatment regimen for EMPD. Although surgical therapies appear to be the preferred treatment for EMPD, conservative approaches such as topical creams can be a viable long-term treatment for select patients versus more radical treatments such as a radical vulvectomy with lymph node dissection. This current study also demonstrates the need for a collaborative effort among institutions to compile their data pertaining to patients presenting with EMPD to increase study sample sizes to explore new treatment options and possibly establish treatment recommendations.