Vulvar Paget's disease: A Single Institution Case-series in Louisiana

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Background

Extramammary Paget's Disease (EMPD) is a rare gynecological adenocarcinoma with a poorly understood pathophysiology affecting older women. Due to its rarity and complexity, there are no 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

- 11 patients diagnosed with vulvar EMPD from January 2015–June 2025 were identified using an electronic medical record database.
- Data 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.

Figure 1: Surgery as Primary Treatment

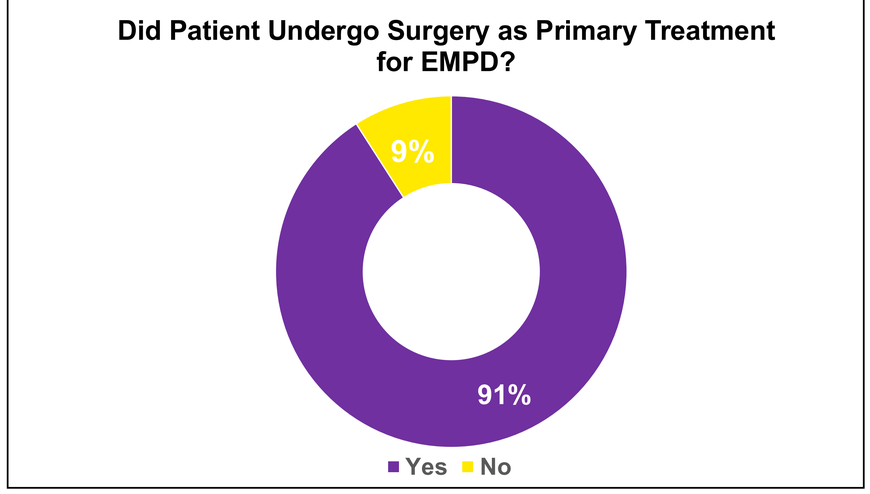


Figure 2: Surgery Type

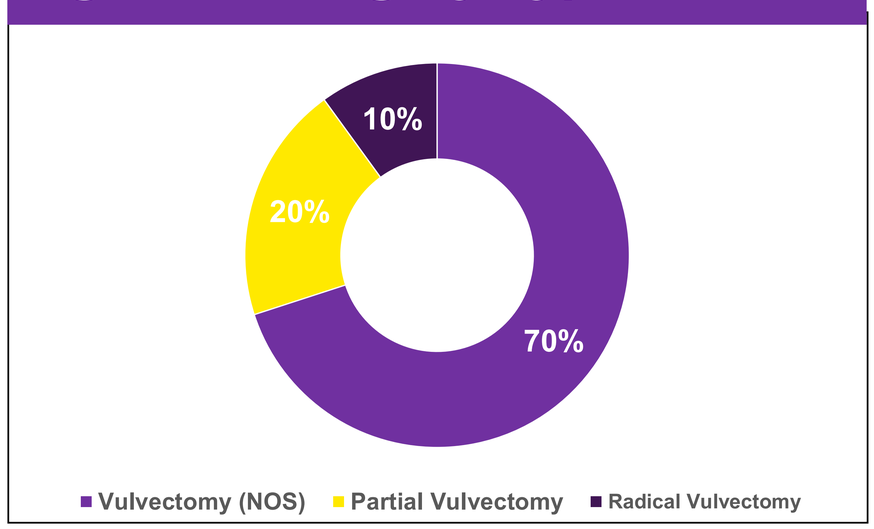
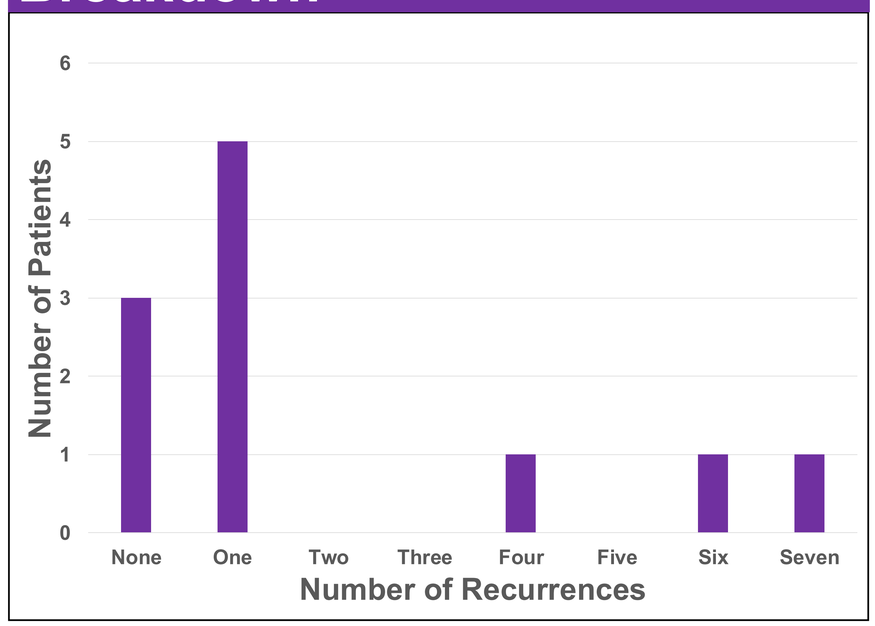


Figure 3: Recurrence Breakdown



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%).
- Eight (73%) experienced a recurrence and all eight had surgery as their primary therapy. 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.

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