

Clinical presentation and Surgical management of Paget's disease of vulva

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Introduction

- Paget's disease of vulva is a rare vulval neoplasm with varied presentation (primary/secondary) (Fig 1)
- Surgical excision is the management for primary disease
- Because of high margin positivity and recurrence rate, many patients undergo multiple excision resulting in significant morbidity.
- Alternative treatment strategies at recurrence are less explored yet.
- The objective of this study was to analyse various clinicopathologic presentations, management and outcomes of women who underwent surgery for Paget's disease.

Methods

The electronic medical record of women who underwent surgery for Paget's disease at our Centre between 2012 and 2023 were reviewed for demographic information, clinical data, pathologic findings, treatment modalities and outcomes. Descriptive statistics were used to summarize results.



Fig.1 Paget's disease of vulva

Results

Nine out of one hundred and sixty-three vulvar surgeries were performed for Paget's disease. The clinico-demographic data is summarised in Table 1. Secondary Paget's disease was ruled out in all of them.

Table 1. Clinico-demographic data of patients

Total number of patients	N = 9
Median age (years)	60 (range 39-69)
Premenopausal	2
Postmenopausal	7
Symptoms	
Itching	7
Redness	5
Itching and vulval redness	3
Leukoplakic lesion	1
Median duration of symptoms (months)	50 (range 12-120)

The histopathology is summarised in Table 2 and the details of treatment at baseline and recurrence are summarised in Table 3.

On a median follow up of 31 months (range 8-66), five recurrences occurred. The median time to recurrence was 14 months (range 10-56)

Table 2 : Histopathology details

Histopathology	N=9
Intraepithelial Paget's disease	4
Invasive adenocarcinoma with Paget's	3
SCC of vulva with coexistent Paget's	2

Table 3: Treatment details at baseline and recurrence

Primary treatment	N=9
Surgery	8
Radical vulvectomy	6
Wide local excision	2
Flap reconstruction	4
Groin node dissection	3
Adjuvant radiotherapy	2
NA-CTRT-debulking surgery	1
Recurrence	N=5
Surgery with flap reconstruction	3
Imiquimod application	1
Chemotherapy	1
Follow up	N=9
Alive with no disease	8
Death	1

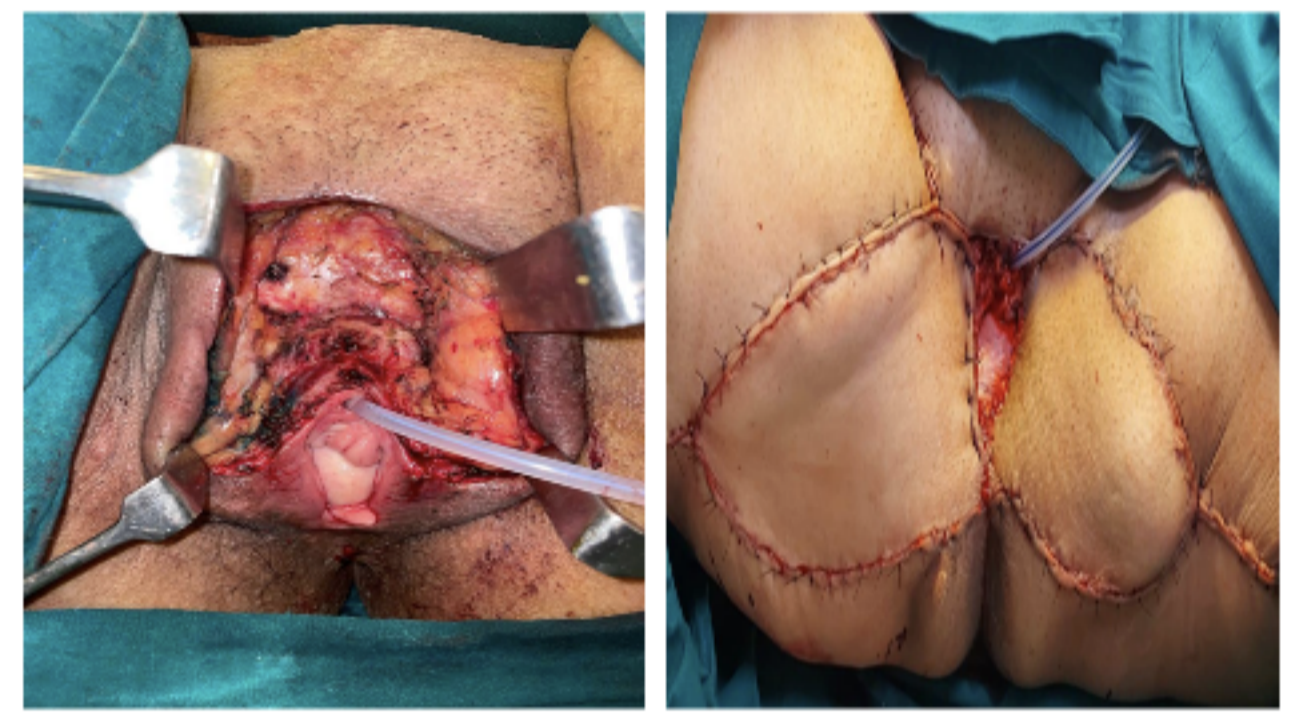


Fig 2. Radical vulvectomy and V-Y plasty

Discussion

- The nonspecific nature of symptoms, social stigma in seeking medical attention, misdiagnosis as dermatoses lead to significant delay in arriving at correct diagnosis and timely management.
- The histological diagnosis is also fraught with difficulties- with invasion, coexistent SCC/VIN often missed in biopsy specimen leading to inadequate primary management.
- Invasion is associated with nodal metastasis. All women with invasive adenocarcinoma had nodal metastasis in this study and required multimodal management.
- Imiquimod has been used for one woman at recurrence of intraepithelial Paget's disease with demonstrated sustained response after 20 weeks of application.

Conclusion

- Clinicians should have a high level of suspicion for vulvar Paget's disease in patients with chronic symptoms and an early biopsy should be performed.
- Longer duration of symptoms prior to presentation could be an attributing factor for some patients presenting in advanced stage.
- Biopsy from multiple sites of a Paget's lesion will help in diagnosing invasive adenocarcinoma or coexisting SCC preoperatively and plan management accordingly
- Regular long term follow up is required in these patients to diagnose recurrence early

Reference

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