

Management of vulval cancer

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1. Scope and background

The purpose of this guideline is to describe the management of histologically confirmed vulval cancer and provide currently available best evidence to health care professionals to provide optimal care for these patients. This guideline also reviews their management options depending on the resources available in the local setting.

Ultimate goal of treating a cancer patient is to cure the disease where possible and to have control of primary disease and delay the recurrences in patients in whom complete cure is not possible. Patients beyond above levels should receive appropriate symptom relieving treatment. Pre-operative staging, individualized treatment planning and appropriate adjuvant treatment and risk based follow up are corner stones in managing these patients.

2. Summary of key recommendations

2.1 Initial assessment

- Patients should undergo speculum and per vaginal examination to assess for involvement of urethra, vagina and anus. Cervix should be inspected for possible synchronous lesions.
- Inguinal area should be examined for lymph node enlargement.

- Wedge or punch biopsy (incisional biopsy) should be taken from the edge of the lesions.
- In instances where excision biopsy is done for small lesions, at least 10mm radical margins should be achieved.
- Chest X ray should be performed in all patients to assess for lung metastasis.
- When available, CT scan of the chest, abdomen and pelvis should be considered in patients with possible stage IB and beyond vulval cancer.
- Full blood count, renal functions, liver functions etc. should be performed to assess the fitness for surgery.
- When possible, multidisciplinary inputs should be obtained before planning surgery for vulval cancer. It is recommended to involve clinicians with oncological background in the decision-making process.

2.2 Management of primary lesion

- Wide local excision with 1 to 2 cm healthy tissues margins is recommended.
- Deep margin is expected to go up to the fascial level.

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2.3 Management of inguinal lymph nodes

- Inguinal lymph node dissection should be done through a separate incision in the groin in contrast to mutilating “butterfly incision” that was practiced in the past.
- Inguinal lymph node dissection should include both superficial and deep groups.
- Inguinal lymph node dissection is indicated in stage IB (>2cm primary tumor or depth of invasion >1mm), II and III disease.
- Lesions within 2 cm of the midline or >4 cm primary lesion require bilateral dissection.

2.4 Adjuvant therapy in vulval cancer

- Post-operative histology review should be done by a senior clinician to decide on need of adjuvant chemo radiotherapy.

2.5 Recurrent vulval cancer

- All patients should undergo biopsy +/- examination under anaesthesia.
- CT scan of chest, abdomen and pelvis is recommended to exclude metastatic disease. When CT scanning is not freely accessible, chest X-ray and ultrasound scan of the abdomen and pelvis should be done.
- Management of recurrent vulval cancer should be planned with multidisciplinary inputs from gynaecological and clinical oncology specialties.
- Radical radiotherapy is an alternative where radical surgical excision is not possible, provided patient has not had radiotherapy before.
- Recommended treatment for groin recurrence is radical excision followed by chemo radiotherapy. When surgical excision is not possible, patient should be referred to a clinical oncologist for primary chemo radiotherapy.

2.6 Management of complications

- Wound infection, wound dehiscence, lymph oedema should be managed in a multidisciplinary setting.

2.7 Follow up

- A follow up based on appropriate history and examination is recommended.

- Follow up frequency should be dictated by risk of recurrence of the cancer.

3. Introduction

Vulval cancer is a relatively rare disease contributing to less than 5% of the gynaecological malignancies. In the past ultra-radical mutilating surgery was the norm in cancer of vulva. But with new knowledge, more conservative surgery is the practice without compromising the survival. New developments such as sentinel lymph node sampling has further reduced the treatment associated morbidity.

4. Recommendations and discussion

4.1 Initial assessment

- Patients should undergo speculum and per vaginal examination to assess for involvement of urethra, vagina and anus. Cervix should be inspected for possible synchronous lesions.
- Inguinal area should be examined for lymph node enlargement.
- Wedge or punch biopsy (incisional biopsy) should be taken from the edge of the lesions.
- In instances where excision biopsy is done for small lesions, at least 10mm radical margins should be achieved.
- Chest X ray should be performed in all patients to assess for lung metastasis.
- When available, CT scan of the chest, abdomen and pelvis should be considered in patients with possible stage IB and beyond vulval cancer.
- Full blood count, renal functions, liver functions etc should be performed to assess the fitness for surgery.
- When possible, multidisciplinary inputs should be obtained before planning surgery for vulval cancer. It is recommended to involve clinicians with oncological background in the decision-making process.

Biopsy from the necrotic center may become falsely negative. Therefore deep peripheral biopsy is recommended. Even if the lesion is small, it is a good practice not to excise the entire lesion at the time of biopsy, as this makes the subsequent definitive surgery difficult to plan. When excision biopsy is performed, radical excision is recommended. CT scan is helpful

in identifying metastatic disease. When CT is not freely available, ultrasound scan of the abdomen/pelvis including the inguinal area should be done. Enlarged inguinal nodes with indeterminate characteristics can be further assessed by fine needle aspiration.

4.2 Management of primary lesion

- Wide local excision with 1 to 2 cm healthy tissues margin is recommended.

- Deep margin is expected to go up to the fascial level.
- Traditionally 1 to 2 cm of healthy tissues are excised with aim for 8 mm pathological clearance¹. However, involvement of vital structures such as anal sphincters, urethra should also be considered when deciding on radicality of excision. However, distal 1 to 2 cm of urethra can be excised without an effect on the continuance¹.

Stage IA	Tumor size less than or equal to 2 cm and stromal invasion is less than or equal to 1mm ³	Wide local excision (WLE) only.
Stage IB	Tumor size more than 2 cm and stromal invasion is more than 1 mm ³	WLE / radical vulvectomy with inguinal lymph node assessment (see below).
Stage II	Tumor of any size with extension to lower one-third of the urethra, lower one-third of the vagina, lower one-third of the anus with negative nodes.	WLE / radical vulvectomy with inguinal lymph node assessment (see below) Excision of the distal urethra and vagina should be considered. When anus is involved, neo adjuvant chemotherapy can be used to reduce the size of the lesion in view of avoiding faecal incontinence. Plastic reconstruction maybe needed in some instances.
Stage III	Tumor of any size with extension to upper part of adjacent perineal structures, or with any number of nonfixed, nonulcerated lymph nodes. Stage IIIA – Tumor of any size with disease extension to upper 2/3rds of the urethra, upper 2/3rds of the vagina, bladder mucosa, rectal mucosa, or regional lymph node metastases less than or equal to 5mm. Stage IIIB – Regional lymph node metastases more than 5mm. Stage IIIC – Regional lymph node metastases with extracapsular spread.	Treat the primary lesion surgically when it is possible. An extenterative type of excision may be needed in some cases. Primary concurrent chemoradiation is an option in this group to avoid significant surgical morbidity. Removal of enlarged inguinal lymph nodes followed by radiotherapy. (See below)

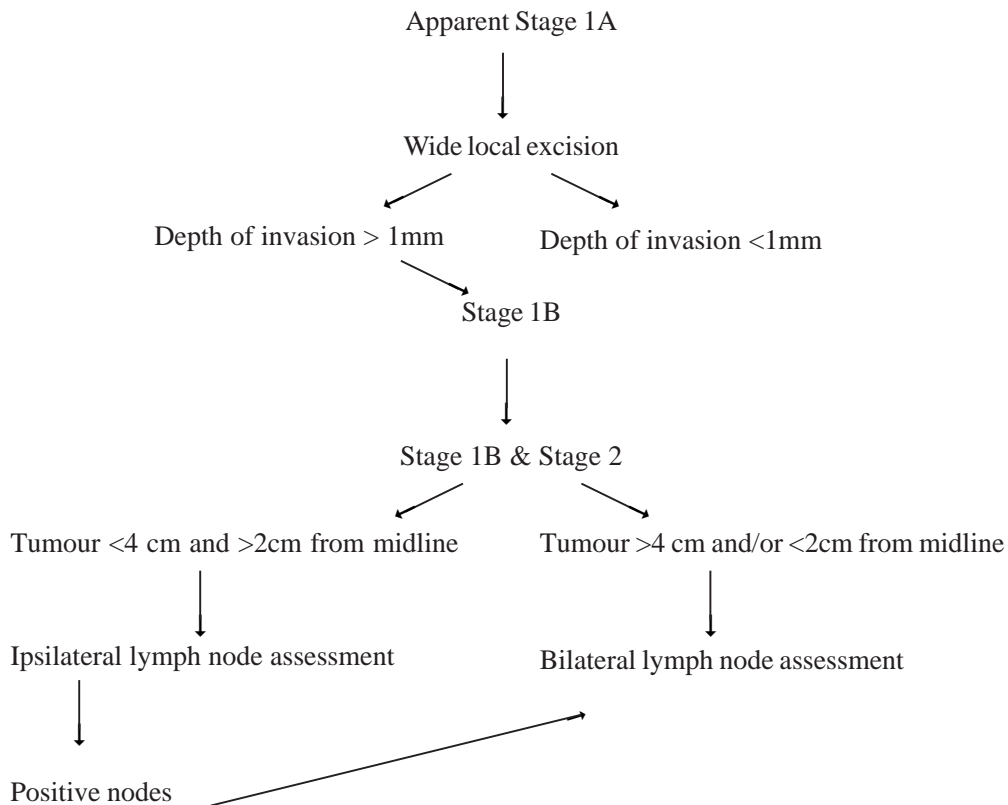
<p>Stage IV</p>	<p>Tumor of any size fixed to bone, or fixed, ulcerated lymph node metastases, or distant metastases.</p> <p>Stage IVA – Disease fixed to pelvic bone, or fixed or ulcerated regional lymph node metastases.</p> <p>Stage IVB – Distant metastases.</p>	<p>No place for radical surgery or radical chemo radiotherapy.</p> <p>Treatment is only in palliatives intent. Early referral to a clinical oncologist.</p>
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4.3 Management of inguinal lymph nodes

- Inguinal lymph node dissection should be done through a separate incision in the groin in contrast to mutilating “butterfly incision” that was practiced in the past.
- Inguinal lymph node dissection should include both superficial and deep groups.
- Inguinal lymph node dissection is indicated in stage IB (> 2cm primarily tumor or depth of invasion >1mm), II and III disease².

- Lesions within 2 cm of the midline or >4 cm primary lesion require bilateral dissection².

Inguinal lymph node assessment is done as a part of the surgical staging. Full dissection should be avoided in patients with clinically positive nodes to avoid severe lymphoedema due to dual treatment. Debulking of enlarged nodes should be followed by chemo radiotherapy. Sentinel lymph node biopsy can be used in lesions <4cm, with no clinical evidence of inguinal lymph node involvement³. This involves injecting a radioactive dye around the lesions followed by imaging with gamma camera. First one or two lymph nodes that receive the injected indicator is excised.



4.4 Adjuvant therapy in vulval cancer

- Post-operative histology review should be done by a senior clinician to decide on need of adjuvant chemo radiotherapy.

Following patients should be referred to a clinical oncologist for adjuvant treatment:

- Positive lymph nodes.
- Involved margins – If possible, these patients should undergo re excision. Alternatively they can be treated with chemo radiotherapy.
- Close margins – Patients with excision margin <5mm should be referred for chemo radiotherapy when re excision is not possible. Close margins are associated with high recurrence rates⁴.

4.5 Recurrent vulval cancer

- All patients should undergo biopsy +/- examination under anaesthesia.
- CT scan of chest, abdomen and pelvis is recommended to exclude metastatic disease. When CT scanning is not freely accessible, Chest X-ray and ultrasound scan of the abdomen and pelvis should be done.
- Management of recurrent vulval cancer should be planned with multi-disciplinary inputs from gynaecological and clinical oncology specialities.
- Radical radiotherapy is an alternative where radical surgical excision is not possible, provided patient has not had radiotherapy before.
- Recommended treatment for groin recurrence is radical excision followed by chemo radiotherapy.

When surgical excision is not possible, patient should be referred to a clinical oncologist for primary chemo radiotherapy.

Approach to local recurrence is similar to management of primary tumour. However, period from the primary treatment, anato-mical distribution of the disease, previous radio therapy as well as physical fitness/performance status are important variables to consider when managing these patients. Previous radical irradiation is associated with cellular changes that prevents the possibility of re-irradiation due to extremely high incidence of organ toxicity (Urinary fistula, Faecal fistula, Skin ulceration, bowel stricture formation, ureteric stricture formation etc).

4.6 Management of complications

- Wound infection, wound dehiscence, lymph oedema should be managed in a multidisciplinary setting.

4.7 Follow up

- A follow up based on appropriate history and examination is recommended.
- Follow up frequency should be dictated by risk of recurrence of the cancer.
- **History:** Ask for perineal itching, soreness, bleeding, discharge or new onset persistent lower limb swelling.
- **Examination:** All patients should undergo examination of the inguinal area for lymphadenopathy and examination of vulva/ vagina.

Follow up interval – risk based approach is recommended

Low risk	Primary unifocal disease treated by surgery only	3-4 monthly clinical assessment for first 2 years. 6 monthly follow up for next 3 years (total 5 years).
High risk	Patients with underlying vulval dermatoses, multifocal/recurrent cancer or Patients who had radiotherapy	3-4 monthly clinical assessment for first 2 years. 6 monthly follow up for next 3 years. Annual follow up there after.

5. Clinical governance

- Patient presenting with suspected vulval cancer should be referred to a specialist gynaecological service within 14 days.
- The definitive treatment of a histologically confirmed case of vulval cancer, should be aimed to occur within 6 weeks from the date of biopsy. Patients should be promptly referred to specialist centers to achieve this target.
- Above time frames are guidance to ensure patient safety and not to be considered as strict rules. Clinical audits on these time frames are highly recommended.

References

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