Management of vulval cancer

Royal College of Obstetricians and Gynaecologists

Setting standards to improve women's health

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CONTENTS

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Review: The RCOG will maintain a watching brief on the need to review the report in the light of new research evidence.

1. INTRODUCTION

This document covers all invasive vulval cancers of any histological type.

As there will be occasions when clinical history and examination alone cannot exclude cancer or preinvasive disease, this document addresses issues of referral for investigation and confirmation, diagnostic procedures and management. The questions of follow-up and outcome assessment are also addressed.

1.1 Objectives

It is hoped that the document will fulfil several objectives:

- to promote a uniformly high standard of care for women with vulval cancer
- to define standard approaches to treatment
- to encourage gynaecological oncologists to develop and participate in clinical trials involving new approaches to management
- to establish means of auditing practice.

This consensus document has been developed by the authors, with input from British Gynaecological Cancer Society members. The authors have also drawn on the experience of previously published guideline materials and other relevant published texts. The document sets out achievable clinical standards and targets for all health professionals involved in the management of patients with vulval malignancy.

1.2 Methodology

Classification of evidence

To ensure that the statements made in this document are evidence based, the current literature was reviewed and critically appraised. The reliability and quality of the evidence given throughout this document has been graded following the NHS Executive classification system, as follows:

Grade A: Based on randomised controlled trials (RCTs).
Grade B: Based on other robust experimental or good observational studies.
Grade C: More limited evidence but the advice relies on expert opinion and has endorsement of respected authorities.

2 SUMMARY OF CONSENSUS STATEMENTS

1 Women with high-grade vulval intraepithelial neoplasia (VIN), VIN with high-grade multicentric disease, VIN in the immunosuppressed and those with Paget's disease or melanoma in situ should be followed up by either specialist multidisciplinary vulval clinics or gynaecological oncologists. [C]

2 In a woman who presents with an unexplained vulval lump, referral should be urgent. [C]

3 In women who present with vulval bleeding, pruritus, pain and ulceration, it is reasonable to use a period of 'treat, watch and wait' as a method of management. This should include the offer of active follow-up until these symptoms resolve or a diagnosis is confirmed. If symptoms persist, referral should be routine, preferably without recourse to biopsy, which may influence subsequent assessment and introduce delay. [C]

4 Vulval cancer should be managed in gynaecological cancer centres/networks by multidisciplinary teams. [B/C]

5 The woman should be seen and managed according to current national directives on waiting times and time from diagnosis to treatment.

6 Radical treatment should not be undertaken without prior biopsy confirmation of malignancy. [C]

7 Wide radical local excision of the primary tumour with a minimum margin of 15 mm of disease-free tissue is often sufficient. [C]

8 Groin node dissection should be omitted in stage 1a squamous cancer, verrucous tumour, basal cell carcinoma and melanoma. [B]

9 Surgery is the cornerstone of therapy for the groin nodes in women with vulval cancer. Individual women who are not fit enough to withstand surgery can be treated with primary radiotherapy. [A/B]

10 Groin node surgery can often be undertaken through separate incisions (triple incision technique) to reduce morbidity. The incidence of skin bridge recurrence in early-stage disease is low. [B]

11 Superficial groin node dissection alone should not be performed, as it is associated with a higher risk of groin node recurrence. [B]

12 In lateral tumours, only an ipsilateral groin node dissection need initially be performed. Contralateral lymphadenectomy may be required if ipsilateral nodes are positive. [B]

13 Preservation of the long saphenous vein may reduce both groin wound and subsequent lower limb problems. [C]

3 BACKGROUND

3.1 Incidence and mortality

Vulval cancer is rare. In the year 2000, there were 996 new cases in the UK, giving a crude incidence rate of 1.7/100000 women (0.8/105 persons) and it was ranked as the 20th most common cancer in women (25th overall).[1] The most recent mortality figures (2002) recorded 364 deaths for all age groups, giving a death rate of 1.2/100 000 women (0.6/105 persons), ranking it the 19th most common cause of cancer death in women (23rd overall).

3.2 Age

Vulval cancer is a disease of elderly women in their 70s and 80s, rarely occurring below the age of 50 years (Figure 1).

Figure 1

Purpose of irradiation	Maximum dose (Gy)
Adjuvant in intent (no macroscopic disease)	45–50 (with no concurrent 5-fluorouracil)
Planned preoperative irradiation	55 (with or without concurrent chemotherapy)
Definitive radical radiotherapy	65 (with concurrent chemotherapy)

3.2 Predisposing factors

There are recognised risks of developing cancer with lichen sclerosus (4 7%),[2] vulval intraepithelial neoplasia (VIN) and multifocal disease (5 90%),[3,4] Paget's disease[5] and melanoma in situ.[6,7] Lichen sclerosus is relatively common and it is not practical to keep all women with the condition on follow-up once their symptoms are controlled. It is recommended, however, that, when the woman is discharged to primary care, written advice is given to her and to her GP, warning that any persistent ulceration or new growth should prompt urgent referral back to the appropriate specialist.[8]

Women with high-grade VIN, VIN with high-grade multicentric disease, VIN in the immunosuppressed and those with Paget's disease or melanoma in situ probably pose higher risks for progression and should be followed up by either gynaecological oncologists or in specialist multidisciplinary vulval clinics.

STATEMENT

Women with high-grade vulval intraepithelial neoplasia (VIN), VIN with high-grade multicentric disease, VIN in the immunosuppressed and those with Paget's disease or melanoma in situ should be followed up by either specialist multidisciplinary vulval clinics or gynaecological oncologists. [C]

3.3 Pathology

Macroscopic

Ninety percent of women at presentation have a visible tumour on clinical examination.

Microscopic

Ninety percent of all vulval cancers are squamous cell carcinomas, with melanoma, Paget's disease, Bartholin's gland tumours, adenocarcinoma and basal cell carcinoma accounting for most of the remaining tumours. The histology is important, as it represents a variable in determining the likelihood of lymph node involvement.

The presence of infiltrative growth patterns, compared with a pushing pattern, is associated with a higher local recurrence rate. Lymphovascular space involvement (LVSI) is also associated with an increased local recurrence rate.[9] LVSI has not been associated with an increased risk of groin node metastasis. Both LVSI and infiltrative growth patterns are markers of poor prognosis but these factors do not indicate the need for adjuvant treatment.

Research is required to establish the influence of these factors on the outcome of this disease.

Spread

Vulval cancer spreads by direct extension to adjacent structures, embolisation to the inguinal and femoral nodes (the regional lymph nodes) or by haematogenous spread. Overall, about 30% of women with operable disease have nodal spread.

Staging

Vulval cancer has been staged surgicopathologically using the International Federation of Gynaecology and Obstetrics (FIGO) staging system since 1994 and has had various modifications, including a subdivision for stage I in 1994 (Appendix 1).

3.6 Prognosis

The 5-year survival in cases with no lymph node involvement is in excess of 80%. This falls to less than 50% if the inguinal nodes are involved and 10–15% if the iliac or other pelvic nodes are involved. A multifactorial analysis of risk factors in squamous vulval cancer demonstrated that nodal status and primary lesion diameter, when considered together, were the only variables associated with prognosis.[10]

3.7 Screening

There is no screening procedure for vulval cancer. Women with carcinoma of the vulva are at an increased risk of developing other genital cancers, particularly cervical cancer. Similarly, women with invasive intraepithelial disease of the cervix are at an increased risk of developing invasive and intraepithelial vulval and vaginal neoplasia.[2]

4 REFERRAL PATHWAYS

Vulval cancer should be managed in gynaecological cancer centres by multidisciplinary teams with specialist nursing expertise. This concept is based upon:

- strong support for centralised referral from a national survey of Scottish gynaecologists.[11]
- indication of more varied practice and worse outcomes from a population-based survey conducted in the West Midlands.[12]
- more favourable outcomes in centres as opposed to peripheral units as documented in a hospital-based study in The Netherlands (this study may be biased because of age differences in the two populations).[13]

4.1 Primary care

The most common presenting symptoms of vulval cancer include pruritus, discomfort or pain, a visible or palpable lesion, ulceration, bleeding dysuria and vaginal discharge.[14,15] In a woman who presents with an unexplained vulval lump, referral should be urgent.[16]

STATEMENT
In a woman who presents with an unexplained vulval lump, referral should be urgent. [C]

In women with vulval bleeding, pruritus, pain and ulceration, it is reasonable to use a period of 'treat, watch and wait' as a method of management. This should include the offer of active follow-up until these symptoms resolve or a diagnosis is confirmed. If symptoms persist, referral should be routine, preferably without recourse to biopsy, which may influence subsequent assessment and introduce delay.[16]

STATEMENT
In women who present with vulval bleeding, pruritus, pain and ulceration, it is reasonable to use a period of 'treat, watch and wait' as a method of management. This should include the offer of active follow-up until these symptoms resolve or a diagnosis is confirmed. If symptoms persist, referral should be routine, preferably without recourse to biopsy, which may influence subsequent assessment and introduce delay. [C]

A postmenopausal woman presenting for the first time with lesions assumed to be vulval condyloma should have histological confirmation of presumed benign pathology. Newly acquired condylomas are unusual in this age group. The woman should be referred to a local unit lead for gynaecological oncology or to the nearest gynaecological cancer centre/network.

4.2 Referral to a gynaecological cancer centre/network

For small suspicious lesions, women should be referred to a gynaecological cancer centre, either after a small biopsy that leaves the lesion identifiable or no biopsy at all. The site and size of the lesion are important variables in treatment planning and if the lesion has been removed prior to referral it might arguably compromise definitive treatment. Ideally, all lesions should be photographed.

Referral should include sending all relevant histopathological material to the specialist gynaecological pathologist in the gynaecological cancer centre.

All new cases of vulval cancer should be discussed at the cancer centre multidisciplinary team meeting. It is also important to identify psychosexual issues prior to treatment.[17]

STATEMENT
Vulval cancer should be managed in gynaecological cancer centres/networks by multidisciplinary teams. [B/C]

The woman should be seen and managed according to current national directives on waiting times and time from diagnosis to treatment.

Results of previous investigations should be available at the visit.

STATEMENT
The woman should be seen and managed according to current national directives on waiting times and time from diagnosis to treatment.

The woman should be informed of the diagnosis and counselled as to the proposed management options and plan, which may include surgery and/or radiotherapy (with or without concurrent chemotherapy).

The appointment should allow for adequate time to counsel and support the woman and, if at all possible, the woman should be introduced to a clinical nurse specialist in gynaecological oncology.

The woman's general practitioner should be informed of the diagnosis and management plan within 2 working days of the clinic visit.

Some women will require an examination under anaesthesia. Such an examination is best performed in the cancer centre or by the gynaecological oncologist who will be responsible for her care. For large cancers of the vulva, it is ideal to have both the gynaecological oncologist and clinical oncologist perform a joint assessment. If a surgical approach is not an option, the woman should be managed by a clinical oncologist with a specific interest in gynaecological malignancy and she should expect to receive an appointment within 2 weeks.

Surgical treatment should commence within 4 weeks of diagnosis. In exceptional circumstances a longer interval may be acceptable, particularly if complex planning is required or significant comorbidities need to be controlled.

Radiotherapy treatment should commence within 4 weeks of the decision to treat by radiotherapy, although in exceptional circumstances a longer interval may be acceptable.

5 DIAGNOSIS AND INVESTIGATIONS

5.1 Examination

When evaluating a vulval lesion, the size and location should be documented. Care should be taken to assess any involvement of the vagina, urethra, base of bladder or anus. With large tumours, one should palpate whether the tumour is infiltrating deep to the pubic and ischial bones. The examination may have to be performed under general anaesthesia because of the pain often associated with large tumours. The presence or absence of groin lymphadenopathy should also be noted.

5.2 Investigation

All diagnoses should be based upon a representative biopsy of the tumour that should include the area of epithelium where there is a transition of normal to malignant tissue. Diagnostic biopsies should be of a sufficient size (greater than 1-mm depth to allow differentiation between superficially invasive and frankly invasive tumours) and orientated to allow quality pathological interpretation. Biopsies should be referred to a pathologist with a specialist interest in gynaecological pathology.

There may be exceptions to these rules. If, for instance an elderly woman with major medical problems and a severely symptomatic lesion presented, a small punch biopsy under local anaesthetic could provide adequate diagnostic information to allow planning of definitive therapy. In certain situations where the clinical diagnosis is apparent and the patient very symptomatic, i.e. heavy bleeding or pain, definitive surgery to the vulval lesion may be performed but biopsy with frozen section is recommended prior to proceeding with any radical procedure.

Although not essential, pre-biopsy photographs are of value in planning treatment, particularly if the diagnostic phase and treatment phases are conducted in separate centres.

After confirming the diagnosis, the objectives of further investigations are to determine the extent of the disease and suitability for treatment. The following investigations are suggested for the majority of patients, although one must accept that, in a predominantly elderly population, this cannot be considered either complete or proscriptive, as each case must be fully assessed according to individual need:

- full blood count (pretreatment assessment)
- biochemical profile (pretreatment assessment, abnormal liver function might suggest metastatic disease)
- chest X-ray (preoperative assessment, exclude metastases)
- electrocardiogram (preoperative assessment)
- cervical smear if normal cervical smear is overdue within the NHS cervical

screening programme
- locally available imaging to assess for concurrent pelvic pathology and retroperitoneal nodes[17–22]
- fine-needle aspiration of any clinically suspicious nodes or other metastases where the result will alter management (i.e. may elect for radiation therapy).

STATEMENT
Radical treatment should not be undertaken without prior biopsy confirmation of malignancy. [C]

6 TREATMENT OF PRIMARY DISEASE

The treatment of vulval cancer is primarily by surgery. This has become more individualised and conservative although the need for adequate resection margins (1 cm) and groin node dissection remain important basic principles. The impetus for more conservative approaches stems from the well-recognised psychosexual sequelae.[23,24] Reconstructive surgery has a role in the management of these cancers. Radiotherapy is used in the adjuvant setting and with or without chemotherapy and surgery in advanced disease.

Management may vary considerably from quite simple to very complex. Each case should be considered on its merits and an agreed plan of management devised by the gynaecological cancer team. Factors such as tumour size, location, medical fitness and the wishes of the patient will all influence management. The management of the nodes and the primary tumour should be considered on their own merits. Tumours should be staged using the FIGO or TNM (tumour, nodes and metastases) classifications. FIGO staging is surgical–pathological and not clinical.

It should be emphasised that these patients are often elderly and have significant comorbidity. As such, they require access to skilled anaesthesia services including an epidural service, high-dependency and or critical care. A key component of patient management is skilled nursing care. All of these services should be available in a cancer centre/network.

6.1 Surgery

Early-stage disease

Depth of invasion
Lesions less than 2 cm in diameter and confined to the vulva or perineum, with stromal invasion less than or equal to 1.0 mm (FIGO Stage Ia) can be managed by wide local excision only, without groin node dissection. This is because the risk of lymph node metastases is negligible.[25]

Dissection of the groin nodes should be performed when the depth of invasion is greater than 1 mm (FIGO Stage 1b or worse) or the maximum diameter of

the tumour is greater than 2 cm (FIGO Stage 2 or worse).[26] This surgery can often be undertaken through separate incisions (triple incision technique) to reduce morbidity. The incidence of skin bridge recurrence in early stage disease is very low.[27]

Surgery to the primary tumour should be radical enough to remove the tumour with adequate margins. The incidence of vulval recurrence has been shown to be related to the measured disease-free surgical margin, as measured in the fixed histopathological specimen. Given the reduction and contraction of tissues following excision and fixation, this equates to at least a 15-mm margin on the fresh surgical specimen. The risk of recurrence increases as the disease-free margins decrease (> 8.0 mm: 0%; 8.0–4.8 mm: 8%; < 4.8 mm: 54%).[9,25] Therefore, wide radical local excision with a minimum margin of 1 cm of disease free tissue should be sufficient.

Excision of atypical skin (lichen sclerosus or VIN) affecting the remainder of the vulva should be considered, as these areas might contain separate foci of invasion. Removal of any lichen sclerosus or VIN skin should not be to the same depth as that for invasive disease unless occult invasion is suspected.

A preoperative vulvoscopy may help in the planning of surgery.

When the surgical margins are found to be less than 1 cm, it may be appropriate to perform a further local resection, although evidence is lacking that this will result in a reduction in local recurrence. There is not enough evidence to recommend adjuvant local therapy routinely in patients with close surgical margins.

STATEMENTS

Wide radical local excision of the primary tumour with a minimum margin of 15 mm of disease-free tissue is often sufficient. [C]

Groin node surgery can often be undertaken through separate incisions (triple incision technique) to reduce morbidity. The incidence of skin bridge recurrence in early-stage disease is low. [B]

Lateral vulval tumours

A lateralised lesion is defined as one in which wide excision, at least 1 cm beyond the visible tumour edge, would not impinge upon a midline structure (clitoris, urethra, vagina, perineal body, anus). Extensive crossover of lymphatic channels of the vulva may result in nodal involvement of the contralateral groins in addition to the ipsilateral groin nodes. Therefore, bilateral groin node dissection is usually required. However, in lateral tumours, only an ipsilateral groin node dissection need initially be performed.28 If the ipsilateral nodes are subsequently shown to be positive for cancer, the contralateral nodes should also be excised or irradiated, as the nodes are more likely to be positive in this scenario.

STATEMENT

In lateral tumours, only an ipsilateral groin node dissection need initially be performed. Contralateral lymphadenectomy may be required if ipsilateral nodes are positive. [B]

Groin node dissection

Appropriate groin node dissection is the single most important factor in decreasing mortality from vulval cancer. However, groin node dissection should be omitted if the patient has Stage 1a disease, as the incidence of lymph node metastases is negligible.[26] It is recommended that the superficial inguinal nodes, as well as the deep femoral nodes, be removed. Superficial inguinal node dissection alone is associated with a higher risk of groin node recurrence.[28] Preservation of the long saphenous vein is reported to reduce both groin wound and subsequent lower limb problems.[29] Following inguinofemoral lymphadenectomy, sartorius muscle transposition may be of benefit in preventing subsequent femoral vessel damage, particularly in those women who are thin and in those in whom adjuvant groin radiation therapy is anticipated.[29] Dye studies and lymphoscintigraphy may be of value in the detection of sentinel nodes,[30–32] although the outcome of this type of intervention is awaiting the outcome of controlled clinical evaluation.

STATEMENTS

Superficial groin node dissection alone should not be performed, as it is associated with a higher risk of groin node recurrence. [B]

Groin node dissection should be omitted in stage 1a squamous cancer, verrucous tumour, basal cell carcinoma and melanoma. [B]

Preservation of the long saphenous vein may reduce both groin wound and subsequent lower limb problems. [C]

Advanced vulval cancer

Surgery to the primary lesion

Resection of advanced disease involves careful preoperative planning and, if reconstruction is required, this should be planned jointly with a plastic surgeon. The size and location of the tumour will influence the surgical approach. Wide, radical, local excision with a minimum of 1 cm disease-free margin may be used but some tumours will require a radical vulvectomy. If these surgical approaches risk sphincter damage leading to urinary or faecal incontinence, treatment by radiotherapy should be considered, either with curative intent or to reduce tumour volume to permit less destructive surgery. Two studies have suggested that preoperative radiation in advanced vulval cancer reduced the need to perform defunctioning stomas.[33,34] It should be noted that, in this post-radiation setting, surgery can be more complicated and there is increased morbidity. Reconstructive surgical techniques should be employed to enable primary surgical closure and to reduce morbidity due to scarring. It should be stressed that the published experience of post-radiation surgery is limited and should not be undertaken lightly. Anovulvectomy might still be considered as an option in selected cases. This is an area where further research is vital.

Management of the groin nodes

Groin node dissection should be undertaken when there are clinically suspicious groin nodes present. In cases with large primary lesions and clinically suspicious nodes, a radical vulvectomy with ‘en bloc’ groin node dissection should be

considered.[27] In cases with fixed or ulcerated groin nodes, surgery and or radiotherapy should be considered. There are no data suggesting the superiority of one treatment over the other, although, if surgery is used, it is likely that postoperative radiation will also be required. Pathological assessment of these nodes should be undertaken prior to radiotherapy, preferably by fine-needle aspiration cytology, in order to maximise the chances of maintaining skin integrity and minimising the risk of wound problems.

Multimodality treatment is increasingly used in the management of advanced vulval cancer to allow for sphincter preserving surgery and as an alternative to surgery for histologically proven involved groin lymph nodes. Surgery following groin radiation may, however, be associated with increased morbidity, both in the groin and in the lower limb. Overall, surgery should still be considered the cornerstone of therapy for the groin nodes.

STATEMENT

Surgery is the cornerstone of therapy for the groin nodes in women with vulval cancer. Individual women who are not fit enough to withstand surgery can be treated with primary radiotherapy. **[A/B]**

Surgical management of nonsquamous vulval cancer

Carcinoma of the Bartholin gland

This is a rare vulval cancer. Histologically, it is usually a squamous carcinoma or adenocarcinoma. The current evidence base is insufficient to suggest different management from squamous tumours. The lesions are often deep-seated or likely to be associated with metastatic disease. The close proximity to the anal sphincter may necessitate partial resection with reconstruction and this may necessitate a defunctioning temporary colostomy.[35,36] Any perimenopausal or postmenopausal woman with a persisting Bartholin abscess or cyst should be suspected of having a possible carcinoma. Appropriate biopsies and histological review should be undertaken.

Basal cell carcinoma and verrucous carcinoma

These squamous variants are rarely associated with lymph node metastases and can be managed by wide local excision. Basal cell carcinomas are also amenable to treatment by radiotherapy, which should be the preferred treatment if resection would compromise function (i.e. would cause sphincter damage).

Malignant melanoma

This group of tumours has not been shown to benefit from block dissection of the groin. Wide local excision is preferred. Relapse in this subgroup is high and closely correlates with the depth of invasion. On the vulva (which includes mucosal surfaces) Breslow's classification[37] is more appropriate than Clarke's levels. As yet, there are no new strategies to minimise the risk of relapse in melanoma.[38]

STATEMENT
Groin node dissection should be omitted in stage 1a squamous cancer, verrucous tumour, basal cell carcinoma and melanoma. [B]

Morbidity related to surgery
The primary objectives of less radical surgery are to reduce morbidity while maintaining high cure rates for early vulval cancers. The complications associated with vulval and inguinal surgery are:

- wound breakdown
- wound infection
- deep vein thrombosis and pulmonary embolism
- pressure sores
- introital stenosis
- urinary incontinence
- rectocele
- faecal incontinence
- inguinal lymphocyst
- lymphoedema
- hernia
- psychosexual complications.

6.2 Radiotherapy

Clinical oncologists supervising treatment should have specific expertise in the management of gynaecological malignancies. They should manage integrated treatment plans involving radiotherapy with or without concurrent chemotherapy.[39]

The factors influencing the need for adjuvant radiotherapy are surgical margins and groin node positivity. There is not enough evidence to recommend adjuvant local therapy routinely in patients with close surgical margins. Adjuvant treatment for positive margins has an improved survival compared with observation alone.[40]

Adjuvant radiotherapy should be considered when either groin has two or more lymph nodes involved with microscopic metastatic disease or there is complete replacement and or extra capsular spread in any node.[41–43] There is no evidence to show whether adjuvant radiotherapy should be given to both sides or to the involved side only. Treatment should be to the groins and the pelvic nodes.

Primary treatment
Radiotherapy, with or without chemotherapy, is increasingly used in the management of advanced vulval cancer. Preoperative radiotherapy may allow for sphincter-preserving surgery. Radiotherapy may also be of use in place of surgery for histologically proven involved groin lymph nodes. It is unknown whether post-radiation groin node removal is advantageous in terms of outcome.

Surgery is the cornerstone of therapy for the groin nodes in women with vulval cancer. Individual women who are not fit enough to withstand surgery can be treated with primary radiotherapy. [A/B]

Treatment schedules

The majority of schedules are based upon those developed by the Toronto Group.[44] Fraction size is important, with 1.7 Gy being close to tolerance, although it is recognised that some centres may use slightly larger fractions (1.8 Gy). Doses will have to be reduced for radical treatment if fractions greater than 1.7 Gy are employed.

Radical treatment will usually require a prophylactic dose (45–50 Gy) to be delivered to the primary and nodal sites and that the tumour is then boosted by a second phase of treatment by electrons, conformal radiotherapy or brachytherapy, to a total dose of 65 Gy. The total prescribed dose is determined by the clinical context (Table 1).

A Cochrane review has suggested that there is no evidence that prophylactic groin irradiation should be used in preference to surgery.[45]

STATEMENT

Surgery is the cornerstone of therapy for the groin nodes in women with vulval cancer. Individual women who are not fit enough to withstand surgery can be treated with primary radiotherapy. [A/B]

6.3 Chemotherapy

The role of chemotherapy in vulval cancer is still debatable but it is increasingly being used in the primary setting. Chemotherapy can be used as an adjuvant postoperatively, concomitant with radiotherapy for node positive disease. This is being evaluated in a number of trials. It is also being used concomitantly with pelvic radiotherapy for the management of inoperable or unresectable disease and it is also being used neoadjuvantly to 'downstage' tumours and to render them more suitable either for delayed primary surgery or radiation therapy. There appears to be no consistent view as to how chemotherapy should best be integrated and many centres have adopted their own policies. A UK consensus would be beneficial.

Chemotherapy has been used for relapsed disease for a number of years. Most recent studies have looked at platinum and 5-fluorouracil-containing regimens or PMB (cisplatin, methotrexate and bleomycin) but mitomycin-C and 5-fluorouracil have also been used. A trial from the European Organisation for Research and Treatment of Cancer in the late 1980s evaluated the use of lomustine (CCNU), methotrexate and bleomycin in locally advanced cases with a surprisingly high activity.[46] Parallels should also be drawn with the management of anal cancer, which has increasingly shifted from being a tumour primarily managed by surgery to a tumour most frequently managed by combination chemotherapy and radiotherapy. Similar discussions are

also continuing in anal cancer as to whether platinum/5-fluorouracil or mitomycin-C/5-fluorouracil is the best schedule and the recent introduction of the 5-fluorouracil pro-drugs capecitabine and Uftoral® (Bristol-Myers-Squibb) – a combination of uracil and tegafur (also known as UFT), which can be given orally, may add to the discussions.

The following scenarios probably best summarise the likely uses of chemotherapy in vulval cancer:

- as a neo-adjuvant to shrink tumour initially considered unresectable
- as a concomitant to radiation for primary management of unresectable tumours
- as a postoperative adjuvant treatment either alone or concomitant to radiation for the management of relapsed disease.

The selection of drugs may be determined by the performance status and age of the patient. Traditionally, many of these tumours arise in elderly women who are often relatively unfit either for radical surgery or for intensive chemotherapy regimens and this may help to determine whether a platinum-based or mitomycin-C-based regimen is selected. There are no randomised trials to support whether one regimen is superior to the other but there is an impression that platinum-based regimens are more likely to be effective. Platinum is the drug of choice in other squamous cancers arising in the cervix, anus, lung, oesophagus, head and neck. Most of the experience will lie with cisplatin and, at the time of writing, the role of carboplatin should be considered as unproven. There are several studies evaluating the use of carboplatin, with or without taxanes, in cervical cancer.

The next debate is whether platinum should be given as a single agent or in combination. There is a body of opinion that would recommend the use of platinum and 5-fluorouracil. This combination has been used effectively as a neoadjuvant in oesophageal and cervical cancers. Those women with primarily unresectable disease, who are of good performance status and with good renal function, should be considered for platinum- and 5-fluorouracil-based regimens or PMB. The use of cisplatin at a dose of 60–80 mg/m^2 every 3 weeks, together with 5-fluorouracil (often given as a protracted infusion over 96 hours) can be recommended, using 750 mg/m^2 per day. The use of a peripherally inserted central catheter (PICC) line is usually advised. Between two and four cycles will be given. Clinical and radiological evaluation should be performed by a multidisciplinary team, to decide whether the tumour has shrunk sufficiently to permit surgery or whether to continue with radiotherapy, either alone or with concomitant cisplatin.

Alternative options would include the use of single-agent cisplatin at a dose of 40–50 mg/m^2 5-fluorouracil would be considered.

6.4 Concomitant chemotherapy and radiotherapy

Chemotherapy used concomitantly with radiation should be considered analogous to use in cervical cancer and either cisplatin alone or cisplatin plus 5-fluorouracil should be considered. If used alone, cisplatin at 40 mg/m^2 weekly, concomitantly with radiotherapy, would be advised. Alternative regimens may include cisplatin and 5-fluorouracil using the regimen above, or PMB given on week-1 and week-4 of a prolonged course of radiation. This should be managed

by a unit experienced in looking after women with vulval cancer, as reactions and toxicity can be quite significant. Women should be referred to their regional centres where gynaecological surgeons and oncologists work closely in teams.

6.5 Postoperative chemotherapy

The use of chemotherapy as a postoperative adjuvant is unproven. Usually, postoperative adjuvant treatment will take the form of radiation therapy. The recent Gynaecological Oncology Group 185 study, which closed prematurely, had tried to examine the role of radiation versus chemo/radiation but failed to achieve the recruitment target. A new international study is under development and is proposing the use of chemo/radiation. If chemotherapy is to be used in the postoperative adjuvant fashion with radiation, it is recommended that cisplatin is used as a single agent, probably at a dose of 40mg/m^2 weekly.

7 TREATMENT OF RECURRENT DISEASE

7.1 Recurrence rates and survival

Recurrence rates for invasive squamous cell carcinoma range from 15% to 33%. In a review of the literature, the vulva was found to be the most common site of recurrence (69.5%) with the groin nodes affected in 24.3%, the pelvis in 15.6% and distant metastases in 18.5%.[47]

Survival following regional relapse is poor and, thus, all attempts to prevent it must be made at the time of primary treatment. Skin-bridge recurrence has been reported to be more likely to occur in patients with positive lymph nodes.[48] If the nodes are known or suspected to be positive at the time of primary treatment, an en-bloc dissection should be considered to remove the tissue between the vulva and involved nodes.

Clinical oncologists and gynaecological surgeons need to work closely together to manage patients with recurrent disease, which can be challenging. Integrating these treatments can, however, be highly rewarding.

Treatment and outcome depend on the site and extent of the recurrence.[47] Wide excision of the local recurrence can result in a 5-year survival rate of 56% when the inguinal nodes are negative.[49] If excision would impair sphincter function, irradiation should be considered as the first choice. If irradiation has already been given to maximum dose, then excision should be considered. Such cases require careful joint planning with clinical oncologists and plastic and reconstructive surgeons experienced in the treatment of vulval disease.

7.2 Groin recurrence

Groin recurrence has a much poorer prognosis and is difficult to manage. In women who have not been treated previously with groin irradiation, radiotherapy (with or without additional surgery) would be the preferred option. The options are much more limited in those who have already been irradiated and palliation, which may include surgery, should be considered. In women who have had both surgery and radiotherapy to the groins, the palliative care team should become involved soon after the confirmation of groin recurrence.

7.3 Chemotherapy for relapsed disease

Chemotherapy for relapsed disease may be determined by what previous treatments have been offered and also by the age and performance status and renal function of the patient. For chemo-naïve patients of good performance status, platinum/5-fluorouracil or PMB are probably the best regimens (Section 6.3). For patients not fit for platinum, mitomycin-C and 5-fluorouracil would be an alternative. Patients with progressive disease or early relapse after a platinum regimen should be considered for phase-2 studies if they are suitable. Drugs likely to be of value in this setting would include the same drugs that would be used in squamous cancers of the cervix, lung, head and neck and oesophagus. Paclitaxel, gemcitabine, vinorelbine, paclitaxel and topotecan would be included in these regimens. A recent EORTC study investigated paclitaxel (as a phase 2 design) in relapsed, pretreated patients and showed reasonable activity. There is some current trial work looking at carboplatin and paclitaxel. However there are no licenses for these drugs and they cannot be used routinely. One problem is that many of these patients are relatively elderly and therefore not good candidates for aggressive novel combinations and this, taken together with their relative rarity, makes clinical trials difficult to perform. Collaboration between the regional UK Centres either through the National Cancer Research Institute or through groups such as the EORTC or the Gynaecological Cancer Group should be encouraged.

8 FOLLOW UP

8.1 Hospital review

As patients who relapse locally have a good chance of cure and/or prolonged remission with prompt re-treatment, the patient should be followed up in an environment where trained personnel are available to recognise the earliest signs or symptoms of recurrence and the morbidity of treatment.

Long-term review is still recommended, as these patients remain at an increased risk of developing carcinomas elsewhere in the genital tract and pelvis. Follow-up intervals are currently arbitrary but the following schedule is suggested:

- 3-monthly first year
- 6-monthly second and third years
- annual review thereafter.

The role of routine hospital follow-up impacting on survival has been questioned for several of the gynaecological tumours. In a retrospective review of 138 women with vulval cancer, routine follow-up did not appear to offer early detection or survival advantage.[50]

8.2 Community care

Patients who have received successful curative therapy will require little community care. Those undergoing palliative or noncurative treatment, who may be symptomatic and expectant of relapse, should receive regular support and surveillance from the general practitioner and community nursing services. Pain control should be monitored regularly. Careful hygiene and dressing of fungating lesions will require very close nursing supervision.

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APPENDIX 1 FIGO Staging

FIGO Stage	Description	TNM stage
	Primary tumour cannot be assessed	TX
	No evidence of primary tumour	T_0
0	Carcinoma in situ, intraepithelial neoplasia Grade III	Tis
I	Tumour confined to vulva or vulva & perineum, 2 cm or less in greatest dimension.	$T_1N_0M_0$
Ia	Tumour confined to vulva or vulva & perineum, ≤ 2 cm in greatest dimension and with stromal invasion ≤ 1 mm.	$T_{1A}N_0M_0$
Ib	Tumour confined to vulva or vulva & perineum, ≤ 2 cm in greatest dimension and with stromal invasion >1 mm.	$T_{1B}N_0M_0$
II	Tumour confined to vulva or vulva & perineum, more than 2 cm in greatest dimension.	$T_2N_0M_0$
III	Tumour invades[a] any of the following: lower urethra, vagina, anus and/or unilateral regional node metastases	$T_1N_1M_0$ $T_2N_1M_0$ $T_3N_0M_0$ $T_3N_1M_0$
IVa	Tumour invades[a] any of the following: bladder mucosa, rectal mucosa, upper urethral mucosa; or is fixed to bone and/or bilateral regional node metastases	$T_1N_2M_0$ $T_2N_2M_0$ $T_3N_2M_0$ T_4 any N M_0
IVb	Any distant metastases, including pelvic lymph nodes	Any T Any N M_1

[a] The depth of invasion is defined as the measurement of the tumour from the epithelial–stromal junction of the adjacent most superficial dermal papilla to the deepest point of invasion

APPENDIX 2 Nursing considerations

Introduction

A patient diagnosed with cancer can expect contact with a variety of professionals and others, in hospital, at home or in a hospice. This includes cancer nurses, palliative care nurses and community nurses as well as other members of the multidisciplinary team.[1] It is self-evident that, at the level of the cancer centre, where the more complex and rarer cancers are treated, appropriately educated specialist nursing staff should manage nursing care. This should include cancer nurse specialists with expertise and experience in the management of patients who have undergone major surgery and radiotherapy with associated change in functional ability, altered body image and altered appearance.

Access to support and information

Within a multidisciplinary team, the clinical nurse specialist is in a key position to be able to address the often complex and sensitive issues identified and experienced by the patient.[2] Similarly, the successful development of medical and nursing partnerships enables women with gynaecological cancers to gain proper access to essential expert knowledge and information and thereby to make informed decisions.[3] The use of clear and accurate written literature should also be promoted. Access to self-help and support groups, such as the Vulval Awareness Campaign Organisation, may also be of significant benefit, allowing women to share experiences and seek support from other women diagnosed and treated for the same condition.

Client group

While a significant proportion of women diagnosed with vulval cancer are over the age of 60 years, there has been an increase in the percentage of younger women diagnosed with vulval malignancies. Consideration should therefore be given to the diversity of the client group and their respective individual needs, (physical, psychological, social, spiritual, emotional), depending on age and circumstances.

Altered body image and psychosexual effects

Vulval cancer management leaves obvious residual effects, and disfigurement and dysfunction will be a part of these women's lives.[3] Access to specialist, trained, psychosexual counsellors and clinical nurse specialists should be available and both women and their partners should be made aware of the potential consequences of treatment during initial consultations prior to therapy (surgery and/or radiotherapy), with reinforcement thereafter.[4] Research indicates that a high proportion of women who have undergone major gynaecological surgery (cervix or vulva) would have liked to have had more information on the after-effects of the operation, including physical, sexual and emotional aspects.[5]

Lymphoedema management

Treatments for vulval cancer frequently involve surgical removal of the inguinofemoral lymph nodes, causing an interruption in the lymphatic pathway.

Additionally, radiotherapy can cause further damage and fibrosis to the lymphatic system.[6,7] The incidence and impact of lower-limb lymphoedema have not been studied widely. One study found that 13 out of 16 women were symptomatic within a 6-week period following groin node dissection for vulval cancer.[8] Other studies suggest that up to 69% of women can be affected.[9,10]

Nursing considerations for women who have undergone treatment for vulval cancer include:

- informing the woman before discharge of the possible risks of developing lower-limb lymphoedema
- educating women on preventative measures that include a meticulous skin regimen, advice on appropriate exercise and movement and the importance of maintaining a healthy body mass index
- ensuring that women are aware of early signs and symptoms associated with the condition
- raising awareness of how to access specialist lymphoedema management for the condition, should it occur.

All women who develop lower-limb lymphoedema should have access to the four cornerstones of lymphoedema care:[11]

- skin care to maintain a good tissue condition and reduce the risk of infection
- external compression in the form of elastic compression garments that help reduce new lymph formation and encourage lymph drainage by improving the efficiency of muscle pump
- a programme of exercise and movement to promote lymph drainage without over exertion
- simple lymphatic drainage: a method of lymph drainage that can be carried out by the patient or carer and involves a series of simple hand movements.

The aims of this regimen are to rehabilitate the cancer patient, to reduce any disability as far as possible, to help the patient to achieve an independent lifestyle and to give the patient the skills to manage their own condition.

Tissue viability

Factors affecting normal tissue viability include surgical intervention, radiation effect and malignant fungating wounds, when disease is advanced or recurred. Most surgical wounds are categorised as acute wounds, healing without complication in an expected time frame.[12] In vulval cancer, however, this is often not the case, as the site of wounds and pressure associated with lymphatic damage can and does lead to wound breakdown and infection, irrespective of all attempts made to minimise it. Therefore, specific wound assessment should highlight the characteristics and nature of the wound.[13] These include:

- location
- size of wound
- presence of slough and/or necrotic tissue
- amount and nature of exudate being produced by the wound
- whether odour is associated with the lesion
- nature and type of pain directly attributed to the wound
- the current state of the skin adjacent to the wound

- the effect on the patient's daily life.

Considerations for wound management include:

- structured tissue viability assessment, incorporating regular review by a tissue viability specialist nurse
- clinical photography
- dietician input and nutritional supplementation
- effective use of appropriate dressings
- responsible antibiotic therapy as and when infection is accurately identified.

The psychological impact of delayed wound healing for the patient must also be recognised and addressed to minimise the risk of longer-term psychological effects, such as depression, social isolation and loss of role in society. Caring for a patient with a fungating lesion when cancer progresses or recurs can offer many challenges. Therefore, it is important to ensure that patients' individual needs and wishes are addressed, to promote optimum quality of life for people with malignant wounds.[14]

Palliative care

Advanced disease presents specific problems associated with the original diagnosis. These include:

- fungating vulval or groin wounds
- progressive lymphoedema
- uncontrollable bleeding
- malodour
- urinary and bowel complications
- pain.

Close collaboration with primary care and palliative care teams to support the patient and their carers during this phase of the cancer experience is an essential element of the service provision of the specialist cancer centre team.

Nursing research

There is still little evidence-based research on which to base clear guidance and protocols of care for women diagnosed with vulval cancer and those who undergo subsequent treatment and care to cure, control or even palliate the disease. With the continued evolution of the National Forum of Gynaecological Oncology Nurses in the United Kingdom and the appointment of a research coordinator to the national committee, consideration should be given to the development and promotion of national multicentre studies into the key aspects of nursing needs of women with vulval cancer.

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APPENDIX 3 Histology: vulval surgical pathology specimens

Clinical information

The clinician should provide an accurate description of the site and appearance of the gross lesion. This can be important if the suspected diagnosis is, for example, carcinoma of the Bartholin gland or verrucous carcinoma. The request should also indicate whether the biopsy was excisional or diagnostic. Ideally, large radical resections should be pinned out on cardboard, kept moist with normal saline and sent as fresh tissue to the pathology department as rapidly as possible. If this is not possible, the specimen should be carefully orientated prior to fixation in the usual way.

Indications for biopsy

- Suspected intraepithelial neoplasia (squamous, Paget's disease, melanocytic)
- Tumour or suspected tumour

Types of specimen

- Punch biopsy
- Small ellipse
- Excision biopsy
- Local/radical excision with or without nodes

Sampling the types of specimen

Punch biopsy

Usually taken in the context of VIN or other dermatological disorder. The size of the biopsy should be recorded. The sample should be examined in its entirety at several levels. Care should be taken to ensure that the sections are cut at right angles to the surface of the skin.

Small ellipse of skin

The biopsy should be measured in three dimensions, care being taken to indicate which is the depth of the specimen. The skin should be cut at right angles to the long axis of the specimen at approximately 2–3 mm intervals. All the tissue should be examined histologically.

Excision biopsy

The tissue should be measured. The size of the lesion and the distance of the lesion from the resection margins should be reported. The sample should be cut so that the pathologist can identify the nature of the lesion, the condition of the adjacent skin, the distance of the tumour and associated intraepithelial neoplasm from the resection margins (lateral and deep).

Local/radical excision with or without lymph nodes

The vulval specimen should be carefully described. Blocks should be taken to identify the type of tumour (or residual tumour), the size and adequacy of excision, the proximity of the resection margins, associated dermatological disease (for example lichen sclerosus) and the presence or absence of lymphovascular permeation.

Reporting the specimens

Punch biopsy

The report should describe:

- the condition of the epidermis (e.g., acanthotic, hyperkeratotic, parakeratotic)
- the presence of features suggesting human papillomavirus infection (e.g. koilocytes, dyskeratotic cells, multinucleated cells)
- the presence or absence of a dermatopathological condition
- the presence or absence of cytological atypia
- the presence or absence of invasive neoplasia.

Cytological atypia should be graded and the report should include a statement saying whether the abnormality is in squamous epithelium, is adenocarcinoma in situ (Paget's disease) or a melanocytic abnormality.

If invasion is present the depth of invasion and the presence or absence of lymphovascular permeation should be stated.

In a punch biopsy it is expected that both intraepithelial and invasive neoplasia will have been incompletely excised.

Small ellipse of skin

The content of the report will be similar to that for a punch biopsy. It may be possible, however, to say whether a focal lesion has been completely excised or not.

Excision biopsy

In the case of an area of intraepithelial neoplasia, the report should confirm the presence of intraepithelial neoplasia, grade it if appropriate, state whether invasion is present or not and say whether the lesion has been completely excised or not.

In the case of a solid neoplasm, the report should:

- confirm the presence of tumour
- identify and grade it
- include its measurements
- state how close it comes to the resection margins (including the deep margin)
- state if it has been completely excised or not
- state if lymphovascular permeation is present or not.

The report should also include a description of the adjacent epidermis and should indicate if any intraepithelial neoplasia has been completely excised or not.

Local/radical excision with or without lymph nodes

In the case of intraepithelial neoplasia, the report should be similar to that for an excision biopsy.

In the case of a solid neoplasm, the report should be similar to that for an excision biopsy.

The lymph nodes should be counted and, unless they are macroscopically infiltrated by tumour, should be cut into slices, which should all be examined histologically. The extent and grade of metastatic disease should be noted; e.g., in the subcapsular sinus only, what percentage of the lymph node/nodes is involved by metastasis and any extracapsular extension.

Histology summary

These follow the known factors that influence both management and outcome.

Primary tumour

1. The total dimensions of the specimen.
2. The dimensions (x 3) of the tumour(s).
3. The total number of tumours.
4. The histological type and grade.
5. Any abnormalities in the adjacent epithelium (i.e. VIN, lichen sclerosus).
6. The maximum depth of invasion.
7. Whether or not there is lymphovascular space involvement.
8. The minimum clearance from all the excision margins.

The nodes

1. The total number of lymph nodes.
2. The presence or absence of tumour in the lymph nodes.
3. The number and site of nodes in which there is tumour.
4. The histological type and grade of tumour in the lymph node(s).
5. Whether the node or nodes are totally replaced by tumour.
6. Whether there is evidence that tumour has breached the node capsule.

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APPENDIX 4 Healthcare professionals involved in the provision of care to women with vulval cancer

- General practitioner
- Gynaecological oncologist
- Gynaecologist (lead cancer clinician in referring cancer unit)
- Clinical oncologist with an interest in gynaecological malignancy
- Medical oncologist
- Specialist gynaecological pathologist
- Clinical nurse specialist in gynaecological oncology
- Lymphoedema specialist
- Psychosexual counsellor
- Clinical psychologist
- Plastic surgeon
- Anaesthetist with an interest in pain management
- Palliative care physician and palliative care teams