Pan Birmingham NHS
Cancer Network

Guideline for the Management of Vulval Cancer

Version History

Version	Date Issued	Brief Summary of Change
2.0	20.02.08	Endorsed by the Governance Committee
2.1	19.11.10	Circulated at NSSG meeting
2.2	13.04.11	With comments from Indy Fernando
2.3	10.05.11	Review by Governance Committee. Endorsed subject to final check by David Luesley
2.4	12.05.11	With comments from David Luesley for final approval by DL.
3.0	18.05.11	Endorsed by Governance Committee

Date Approved by Network Governance	**May 2011**

Date for Review	**May 2014**

Changes Made During Review in 2011

- *In the 'radiotherapy and chemo-radiation' section, "those patients unfit for radical treatment maybe given palliative radiotherapy: having doses of 20 Gy in 5 fractions to 30 Gy in 10 fractions " was added.*
- *In the 'recurrent disease' - "palliative radiation maybe used in suitable cases at a dose of 20 Gy in 5 fractions or 30 Gy in 10 fractions can be used, particularly in patients who have advanced metastatic disease" was added.*

1 Scope of the Guideline

This guidance has been produced to support the following:

- The management of patients with suspected vulval cancer
- The management of patients diagnosed with vulval cancer

2 Guideline Background

All Trusts undertaking gynaecological surgery in the Pan Birmingham Cancer Network are recognised as cancer units. One Trust (Sandwell and West Birmingham NHS Trust, City Hospital Site) is recognised as the gynaecological cancer centre. The objectives are in line with national guidance in that all women with vulval cancer will be managed within the gynaecological cancer centre.

Guideline Statements

3 Referral and Diagnosis

3.1 All patients with a lesion suspicious of vulval cancer on clinical examination should be referred to a local gynaecological multi disciplinary team and seen within 2 weeks: 90% of vulval cancers will have a visible lesion, lump, ulcer or raised irregular epithelium. Discomfort and irritation are very common. Most cases occur in women over the age of 65 years.

3.2 If cancer of any type is suspected, patients should be referred directly to the gynaecological cancer centre. If there is doubt, a diagnostic biopsy should be taken. Excisional biopsies should be avoided as treatment planning depends upon the location and size of the tumour help to inform the extent of excision and the need for ipsilateral or bilateral node dissection. **Total removal of a lesion prior to referral may compromise the subsequent use of sentinel node imaging techniques.**

3.3 The diagnosis of vulval cancer should usually be confirmed by biopsy prior to definitive surgery (a representative biopsy of the tumour that should include the area of epithelium where there is a transition of normal to malignant tissue). In situations where the clinical diagnosis is apparent and the patient severely symptomatic, surgery to the vulval lesion may be performed without prior biopsies. Surgery to the groin nodes should only be performed at this time if there is strong clinical suspicion of nodal disease or there is cytology or histological confirmation of nodal involvement.

3.4 Patients diagnosed with vulval cancer should be referred to the gynaecological cancer centre for management. All relevant histological material should be sent to the specialist gynaecological pathologist at the time of referral.

4 Staging and Primary Treatment

4.1 One staging system applies to vulval cancer. In the UK FIGO[1] staging is used in preference (see appendix 1).

4.2 Basal cell carcinoma or superficially invasive squamous (stage 1a) carcinoma

Patients diagnosed with basal cell carcinoma or superficially invasive squamous (stage 1a) carcinoma rarely if ever have lymph node metastases. They are managed by wide local excision. Wide local excision is a term used to define an excisional procedure that aims to provide a margin of 10mm of tumour free surrounding tissue, after fixation, on all aspects of the tumour. Generally the depth should be as deep as Colle's fascia.

4.3 Malignant Melanoma

4.3.1 Patients diagnosed with malignant melanoma may have lymph node metastases. There is no evidence that resecting nodes routinely improves outcomes. It is unknown if patients' benefit from excising enlarged and or suspicious lymph nodes. Grossly involved groin nodes should be resected en-bloc with the primary vulval lesion.

4.3.2 Investigations for Patients with Melanoma

a) No investigations are necessary for patients with stage I disease. Stage I and IIA melanoma patients should not be staged by imaging, as the true-positive pick-up rate is low and the false-positive rate is high (Roberts et al 2002).[3.]
b) Patients at intermediate or high risk of recurrent disease (stage IIB and over) should have the following staging investigations: chest x-ray; liver ultrasound or computed tomographic (CT) scan with contrast of chest, abdomen and pelvis (Roberts et al 2002). [3.]

4.3.3 Patients with melanoma should also be discussed at the relevant skin MDT.

4.4 Squamous Tumours >Stage IA, Adenocarcinomas or Sarcomas

4.4.1 Patients diagnosed with squamous tumours >Stage IA, adenocarcinomas or sarcomas may have lymph node metastases. Patients with stage I or II tumours without clinical evidence of nodal involvement should be offered sentinel node imaging. All other stages of vulval cancer and those with clinically involved nodes may not benefit from this technique (there are not supportive published data available). If either sentinel node is positive patients should be offered further surgery to remove all nodal tissue in the groin or adjuvant radiotherapy to both groins and the pelvis.

4.4.2 Patients with sarcoma should also be discussed at the sarcoma MDT.

4.5 Groin Node Involvement

4.5.1 If there is evidence of groin node involvement, MRI scanning should be performed prior to surgery and scans should include the pelvic nodes. The basic guiding principles are:

a) Lateralised tumours uncommonly involve the ipsilateral nodes
b) Stage I and probably stage II tumours can be managed by separate groin incisions or sentinel node imaging and selective resection
c) Suspicious lymph nodes should be managed en-bloc with the primary lesion
d) If primary lesions are large and their excision might compromise sphincter function, primary radiation, chemoradiation or neoadjuvant chemotherapy should be considered
e) Surgery should be considered in all cases where radiation, chemoradiation or chemotherapy has resulted in an incomplete response. If there has been a complete response multiple biopsies should be considered to confirm complete remission
f) In patients presenting with enlarged, fixed and or ulcerated groin lymph nodes, biopsy or fine needle aspiration should be considered prior to initial treatment with radiation or chemoradiation

4.5.2 A lateralised lesion is defined as one in which wide local excision (allowing a margin of at least 1cm of normal tissue after fixation) would not impinge upon a midline structure such as the clitoris, urethra or anus.

4.5.3 The extent of radical excision depends upon the size and site of the primary tumour. The principle remains one in which the intent is to excise all suspicious epithelium with a minimum margin of 1cm normal tissue after fixation. For a large central lesion this could entail classical radical vulvectomy, for a small lateral lesion, wide local excision.

5 Radiotherapy and Chemoradiation

5.1. Patients undergoing adjuvant treatment should be treated with 45 Gy to 50 Gy only. Some patients with extensive nodal disease where there is a high risk, particularly of local and metastatic recurrence, can be considered for chemo-radiation. This will be discussed at the multidisciplinary team meeting on an individual patient basis.

5.2 Patients undergoing neo-adjuvant pre-operative radiation can be treated to a maximum of 55 Gy to 65 Gy. Chemo-radiation is appropriate for these patients if they are suitably fit.

5.3 Patients undergoing definitive chemo-radiation should receive a maximum of 65 Gy with concurrent 5FU +/- mitomycin C or cisplatinum in a 2 phase treatment. The radiotherapy fraction size is 1.8 Gy if the treatment is given concurrently with the chemotherapy. The chemotherapy regimes used are 5FU alone, mitomycin C/5FU, or cisplatin +/- 5FU.

5.4 Those patients unfit for radical treatment maybe given palliative radiotherapy: doses of 20 Gy in 5 fractions to 30 Gy in 10 fractions.

6 Recurrent Disease

6.1 In local recurrence irradiation should be the first choice if the excision would impair sphincter function. If irradiation has already been given to maximum dose, then excision should be considered.

6.2 Groin recurrence is much more difficult to manage. In patients who have not received radiation and who have histologically confirmed recurrence, radiotherapy should be performed first. Resection should be considered if the response to radiotherapy is not complete. Patients who have been irradiated should be offered palliative resection if possible.

6.3 Groin recurrence management should be discussed on an individual basis. These tumours are rare and isolated groin node recurrences are also infrequent. For example, if the patient has had a long disease free interval, they are unlikely to have problems with wound healing and then operable groin recurrences are probably best treated initially with surgery followed by post-operative radiotherapy. High doses of radiation on the other hand in that situation would probably lead to excessive fibrosis and lymphoedema. However, if there is concern about wound healing following surgery or the recurrence is of borderline operability, then it seems reasonable to offer radiation first.

6.4 Palliative radiation may be used in suitable cases at a dose of 20 Gy in 5 fractions or 30 Gy in 10 fractions can be used, particularly in patients who have advanced metastatic disesae.

7 Follow-Up

Please see the guidelines for the follow-up of gynaecological cancers at:
http://www.birminghamcancer.nhs.uk/staff/clinical-guidelines/gynaecology

8 Patient Information and Counselling

8.1 All patients, and with their consent, their partners will be given access to appropriate written information during their investigation and treatment, and on diagnosis will be given the opportunity to discuss their management with a clinical nurse specialist who is a member of the relevant multi disciplinary team . The patient should have a method of access to the gynaecological team at all times.

8.2 Access to psychological support will be available if required. All patients should undergo an Holistic Needs Assessment and onward referral as required.

9 Clinical Trials

9.1 Wherever possible, patients who are eligible should be offered the opportunity to participate in National Institute for Health Research portfolio clinical trials and other well designed studies.

9.2 Where a study is only open at one Trust in the Network, patients should be referred for trial entry. A list of studies available at each Trust is available from Pan Birmingham Cancer Research Network. Email: PBCRN@westmidlands.nhs.uk.

9.3 Patients who have been recruited into a clinical trial will be followed up as defined in the protocol.

Monitoring of the Guideline

Compliance with the guidance will be reviewed in 2011 and considered as a topic for audit by the NSSG in 2011\2012.

References

1. The International Federation of Gynaecology and Obstetrics (**FIGO**), 2000. **Staging classifications and clinical practice guidelines of gynaecologic cancers** (pages 6-25). http://www.figo.org
2. The Royal College of Obstetricians and Gynaecologists, 1999. **Clinical Recommendations for the Management of Vulval Cancer** RCOG, London.
3. Roberts et al 2002. UK Guidelines for the management of cutaneous melanoma. **British Journal of Dermatology** 146 7-17.

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Approval Date of Network Site Specific Group: Date May 2011

Date Approval by the Clinical Governance Team: Date May 2011

Approval Signatures

Pan Birmingham Cancer Network Governance Committee Chair

Name: Doug Wulff

Signature: Date May 2011

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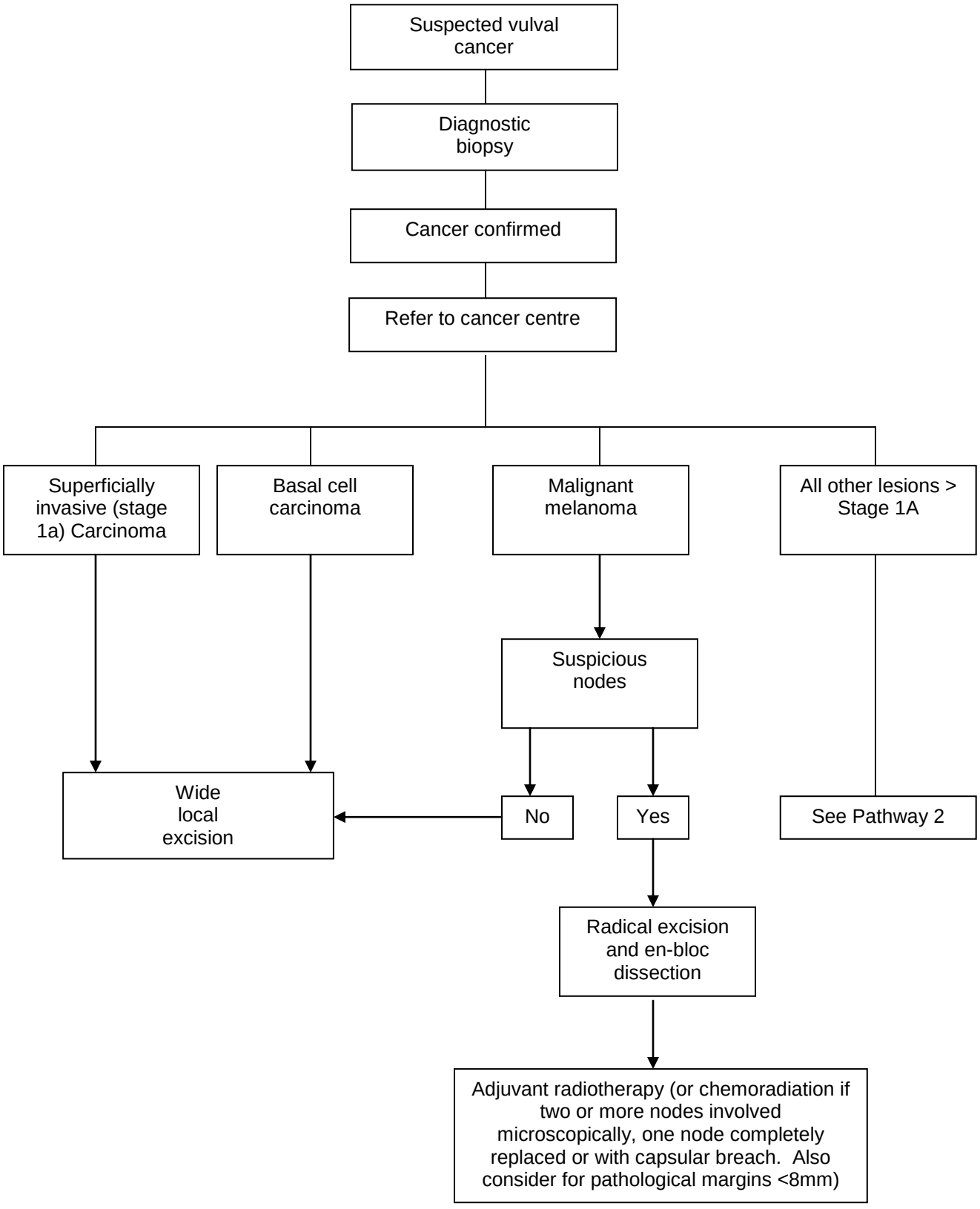
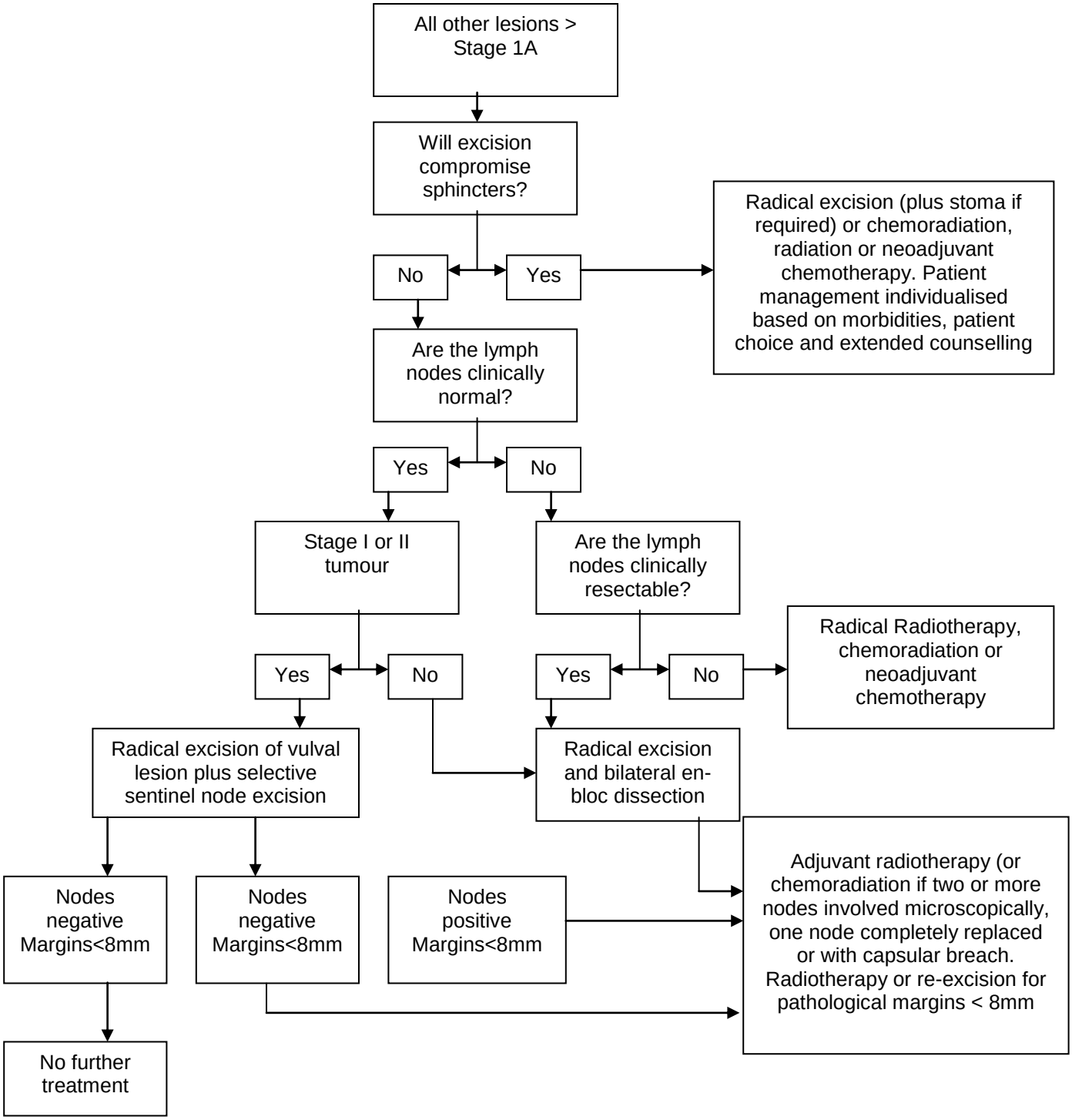
Figure 1 - Patient Pathway for Vulval Cancer (1)



ENDORSED BY THE GOVERNANCE COMMITTEE

Figure 2 - Patient Pathway for Vulval Cancer (2)



Appendix 1: Staging of Vulval Cancer

FIGO	Description
Stage I	**Tumour confined to the Vulva**
Stage Ia	Lesions ≤2cm in size, confined to the vulva or perineum and with stromal invasion ≤1mm. No nodal metastasis
Stage Ib	Lesions >2cm in size or with stromal invasion >1mm confined to the vulva or perineum. No nodal metastasis
Stage II	**Tumour of any size with extension to adjacent perineal structures (Lower 1/3 urethra; Lower 1/3 vagina; Anus) with negative nodes**
Stage III	**Tumour of any size with or without extension to adjacent perineal structures (Lower 1/3 urethra; Lower 1/3 vagina; Anus) with positive inguinofemoral nodes**
IIIa	(i) With 1 lymph node metastasis (≥ 5mm), or (ii) 1-2 lymph node metastasis(es) (<5mm)
IIIb	(i) With 2 or more lymph node metastases (≥ 5mm), or (ii) 3 or more lymph node metastases (<5mm)
IIIc	With positive nodes with extracapsular spread
Stage IV	**Tumour invades other regional (upper 2/3 urethra;2/3 Vagina) or distant structures**
IVa	Tumour invades any of the following (i) Upper urethral and or vaginal mucosa;bladder mucosa; rectal mucosa or fixed to pelvic bone, or (ii) Fixed or ulcerated inguinofemoral lymph nodes.
IVb	Any distant metastasis including pelvic lymph nodes