the eve appeal

An overview of cancer of the vulva

Guidance for GP and community nurses

Contents

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1059
women diagnosed in 2016*
(*ONS 2018)

60%
of women were unable to correctly identify the vulva on an anatomical diagram

Often affects older women but women are being diagnosed at a younger age:
under 45: n=56
45-59; n=215
60-74: n=345
over 75: n=443

Age standardised 5-year survival is
67.9%
and a predicted 10-year survival of
56.6%
in all age groups.

Office for National Statistics (ONS), (2018) Cancer registration statistics, England. Retrieved from ONS: https://www.ons.gov.uk/peoplepopulationandcommunity/healthandsocialcare/conditionsanddiseases/datasets/cancerregistrationstatistics-cancerregistrationstatisticsengland

Introduction

This module provides an overview of vulval cancer to nurses and other HCPs who are new to the field of gynaecology or those interested in developing their knowledge of the disease and its affect on women. It would be especially helpful for those in primary care to promote early diagnosis and management of long term effects.

No preparation or learning materials are required for this learning resource, will require online access. It should take approximately one hour to complete including reflection time.

Gynaecological cancers

There are five cancers that affect the female reproductive system, these are known as gynaecological cancers. Cancer of the vulva (vulval/vulvar) relates to disease that starts within the external female genitals. It is a rare cancer that makes up one of the group of gynaecological cancers.

There are different types of vulval cancer all of which are invasive and malignant lesions that invade either the skin or more rarely the glands of the vulva. Some of these cancers may have developed from pre-invasive conditions such as vulvar intraepithelial neoplasia (VIN) or lichen sclerosus. Vulval cancer is associated with key signs and symptoms that act as red flags for suspecting cancer. Early detection influences treatment options and survival benefit. Treatment pathways vary with the mainstay of treatment being surgery, however radiotherapy and chemotherapy may also be required.

Of those diagnosed and treated over 53% will survive 10 or more years following treatment. Those women may develop late effects and consequences of treatment. Primary care nurses are well placed to influence practice at each stage of the patient's journey from prevention, early detection through to treatment and survivorship.

Aims

To identify & build on your existing knowledge & skills in relation to vulval cancer and to enable you to feel confident in meeting the needs of women in your care.

Learning outcomes

1. Identify key anatomical structures of the vulva/correctly label a diagram of the vulva
2. Identify symptoms that may raise concern for vulval cancer and lead to a diagnosis.
3. Describe treatment(s) used to treat vulval cancer.
4. Discuss the impact on the physiological, psychological and social well-being of women living with and beyond vulval cancer and on their partners.
5. To consider how you can influence the care of women with vulval cancer within your clinical practice.

Test existing knowledge

See case study (on next page), reflect on the study and consider:

- If this lady presented in your work place, what skills and knowledge do you already have and what do you need to develop to enable you to support her?
- What do you think this woman's care needs are from a physical, psychological & social aspect?

'Vulvar cancer is associated with key signs and symptoms that act as red flags for suspecting cancer.'

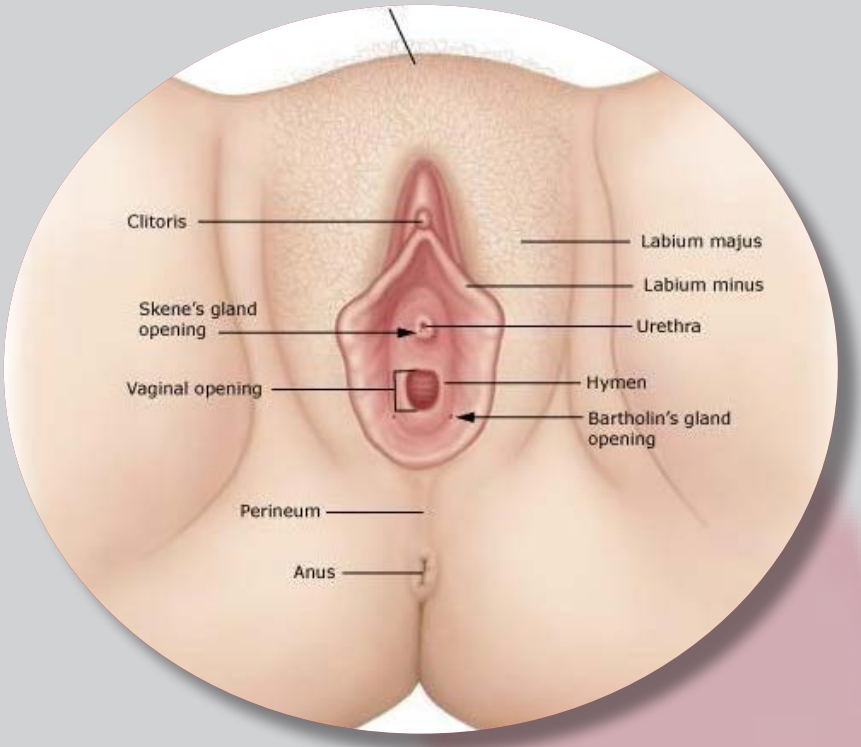
Case study

Carol is 50 years old, she is married to John and they have two children Sam 18 and Ali 16. Carol worked full time but took early retirement on ill health grounds. Carol has been treated for vulval cancer. She was diagnosed when she was 40 years old. Consider her history and when you could have made any interventions, reflect on her story and what you think her care needs may have been. Think about your practice and if Carol's story could influence it.

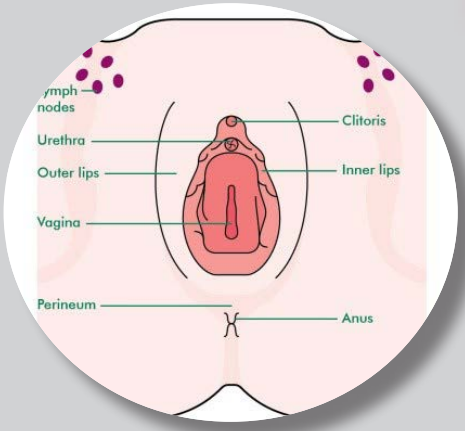
2008	• 2 week wait referral for suspected cancer of the vulva with suspicious lesion on vulva. Biopsy confirmed well differentiated squamous cell cancer, 2cm lesion on left labia not near clitoris or urethra. • Left hemi-vulvectomy and left groin node dissection. • Stage 2 vulval cancer – for adjuvant external beam radiotherapy (25 sessions).
2009	• Vulval itching - ?thrush. Biopsy ruled out recurrence. • Developed lymphoedema – pubic & left thigh – supplied compression shorts.
2010	• Lichen sclerosis on right side of vulva. • Biopsy showed chronic inflammation – treated with Dermovate. • Surgery to remove lichen sclerosis – VIN I (low grade)
2011	• Lymphoedema. Dyspareunia (painful sex). Dysmenorrhea (painful periods) • Further EUA, hysteroscopy and insertion of coil. • Skin changes – inflammation, hyperkeratosis, & radiotherapy changes telangiectasia. • HNA – Dyspareunia, urinary incontinence (since RT but first time of reporting) • Ref to continence service/uro-gynae.
2012	• New symptoms – vulval pain and tear in perineum that wouldn't heal. • EUA & biopsy • Left sided recurrence, right side lichen sclerosis and invasive vulval cancer. • Surgery – radical vulvectomy, reconstruction, lotus petal flap and right groin node dissection. • Wound healing difficulties, MRSA & cellulitis. • EUA – VIN I, II, III
2013	• Vaginal discharge, polymenorrhagia • EUA, excision of vulval lesion, skinning vulvectomy & biopsy. VIN III (high grade)
2014	• Wide excision posterior fourchette & reconstruction flap. • Recurrent squamous cell cancer. • Exenteration – laparotomy, ano-vulvectomy, closure of bladder insertion of suprapubic catheter, AP resection, stoma formation, total abdominal hysterectomy, bilateral saplingo-oopherectomy, VRAM flap (surgery for curative intent)
2016	• Reduction of VRAM flap by plastic surgeon. • Permanent catheter. • Stoma OK • Menopause symptoms – HRT
2018	• Remains on yearly follow up with MRI

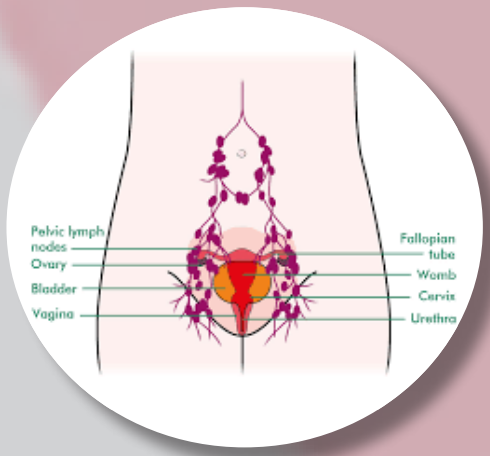
Anatomy of the vulva

The main structures of the vulva are: Mons pubis; Labial minora; Labia majora; Clitoris; Urethral meatus; Introitus/vaginal opening; Bartholin glands; Perineum.



Knowledge of the lymphatic system to the vulva is also important as this will influence staging and treatment options. Inguinal or femoral (groin) nodes.





Signs and symptoms

- Vulval itching that lasts more than one month
- A cut or sore on the vulva that won't heal
- A lump or mass on the vulva
- Unexplained vulval pain
- Bleeding or blood-stained discharge
- Burning when passing urine
- Any change in size, colour or texture of a birthmark or mole in the vulval area

'Knowledge of the lymphatic system to the vulva is also important.'

Risk factors

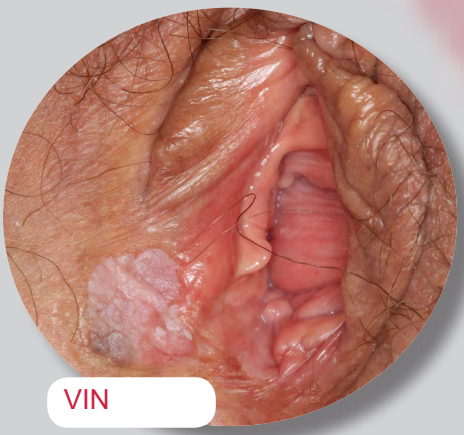
- Age (more common in women over 65yrs old)
- Vulval intraepithelial neoplasia (VIN)
- Lichen sclerosus
- Paget's disease
- HPV – human papillomavirus
- Other STI's – herpes, HIV
- Smoking
- Other health conditions eg systemic lupus erythematosus

Differential diagnosis

Remember that cancer of the vulva is very rare and there are various other benign conditions that should be considered as differential diagnosis. Examination should be considered in line with other risk factors. (WARNING: Please be aware that the pictures are graphic).

- **Vulval intraepithelial neoplasia (VIN)**
- **Vulval warts**
- **Paget's disease**
- **Lichen sclerosis**
- Lichen planus, papulosquamous form
- Lichen simplex
- Major aphthous ulcers

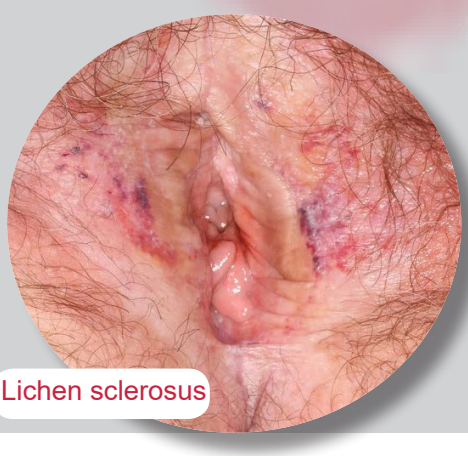
Those in bold are pictured below, as they are the most common.



VIN

Vulval warts

Paget's disease



Lichen sclerosus

Types of cancer - histology

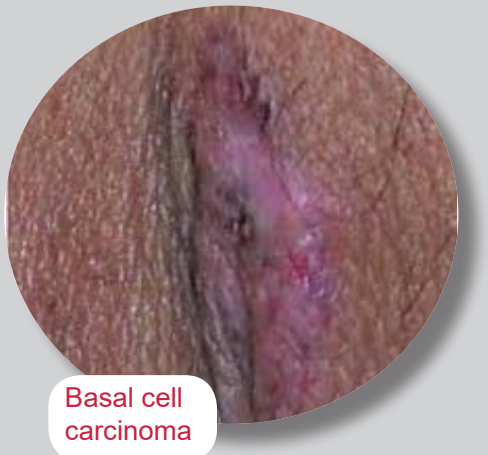
Squamous cell carcinoma: Most vulval cancers (90 per cent) develop from squamous cells, the skin cells of the vulva. These cancers usually grow very slowly over a few years.

Squamous cell carcinoma

Melanoma: Vulval melanomas develop from the melanocytes, the cells that produce melanin and give skin its colour. Only about 2 to 4 per cent of vulval cancers are melanoma.

Melanoma

Basal cell carcinoma: Although slow-growing, the vulvar basal cell carcinoma is a malignant tumour, and is capable of metastasizing to the lymph nodes of the groin.

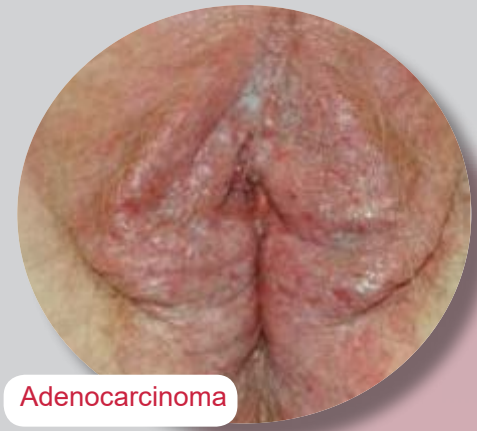


Basal cell carcinoma

‘Cancer of the vulva is very rare.’

Types of cancer - histology

Adenocarcinoma: These are very rare. They develop from cells that line glands in the vulval skin. Paget's disease of the vulva is a pre-malignant condition where glandular cells spread outwards and across the vulval skin.



Adenocarcinoma

Verrucous carcinoma: This rare, very slow-growing type of cancer looks like a large wart. Sarcomas: These are extremely rare. Sarcomas develop from cells in tissue, such as muscle or fat under the skin, and tend to grow more quickly than other types of cancer.

Verrucous carcinoma

Check learning so far

Which of these patients would you consider most likely to have cancer of the vulva.

A. 35 year old woman, long history of abnormal cervical smears, attending for routine smear (annual due to previous CIN). Complains of vulval irritation and itching, on examination lesion superficial lesion on left labia.

a. Consider VIN initially due to history of abnormal smears, this would suggest HPV present. Consider referral to vulvoscopy clinic.

B. 72 year old, complains of 3 month history recurrent UTI symptoms of difficulty in passing urine and pain, but urine samples negative. On examination 3cm warty looking lesion on anterior vulva located between clitoris and urethral meatus.

a. Likely malignancy due to age, location of lesion, unusual for woman of this age to develop warts.

C. 48 year old, attended with changes to appearance of vulva, dark coloured lesion, skin intact, notice lesion 6 months ago but now increasing in size.

a. Consider malignant melanoma – would need referral for review.

Diagnosis

To diagnose vulval cancer a tissue biopsy is required. Some women may undergo imaging to help with staging of the disease but often staging is confirmed following surgery.

Staging:

STAGE 1 Tumour confined to the vulva, with negative node biopsy.

STAGE 2 Tumour of any size with extension to adjacent perineal structures (lower third of urethra, vagina or anus) with negative nodes.

STAGE 3 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.

STAGE 4 Tumour invades other regional (upper 2/3 urethra; 2/3 vagina) or distant structures.

(Staged using International Federation of Gynaecology and Obstetrics – FIGO staging)

Survival (5 year) in disease which has not spread to lymph nodes is over

80%

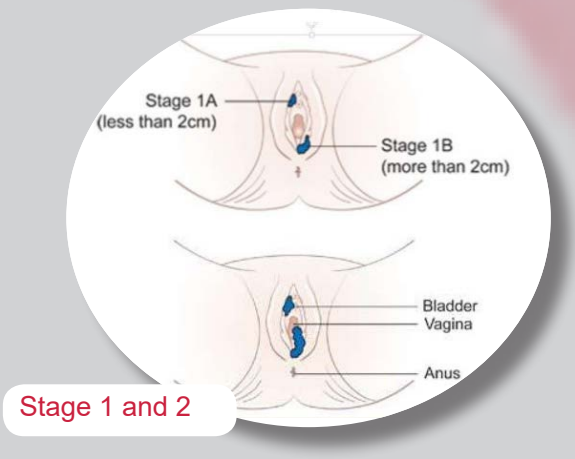
Survival (5 year) is less than

50%

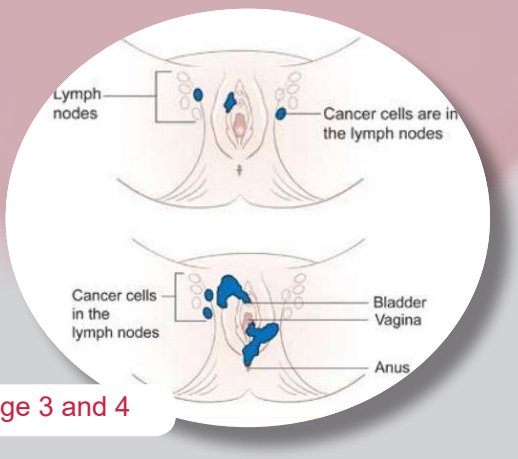
if inguinal nodes are involved and only

10-15%

if other nodes are involved.



Stage 1 and 2



Stage 3 and 4

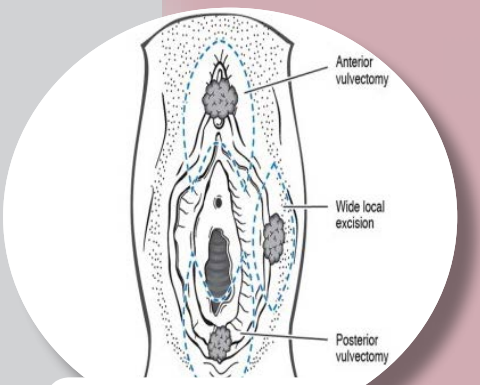
‘Survival (5 year) is less than 50% is inguinal nodes are affected.’

Treatments

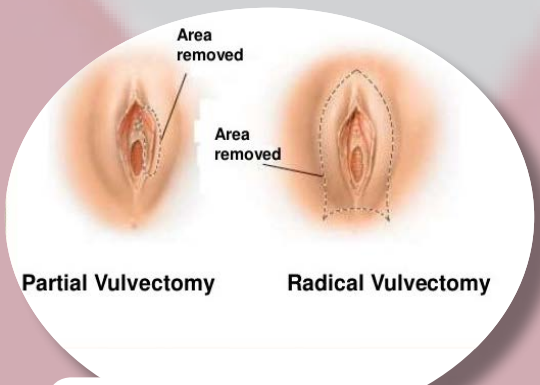
The main treatment is surgery, with the aim to remove the cancer with a 1cm margin of healthy (non-cancerous) tissue. Surgery may also include groin node dissection and if necessary reconstruction surgery.

The main surgeries are :

- Wide local excision,
- Anterior or posterior vulvectomy,
- Unilateral vulvectomy,
- Total vulvectomy,
- Exenteration.

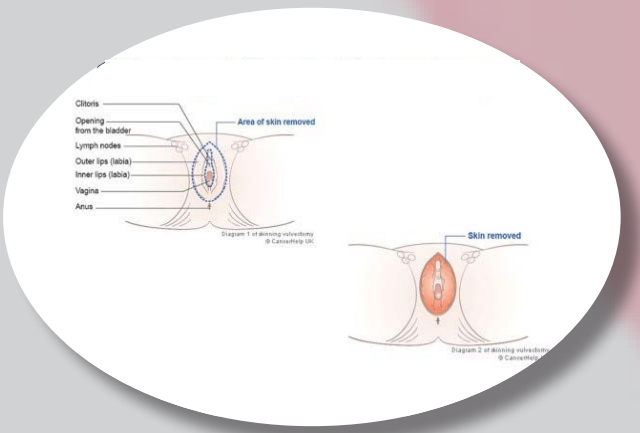


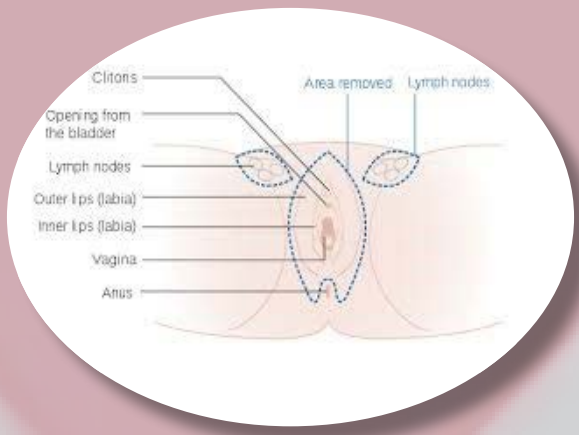
Vulvectomy

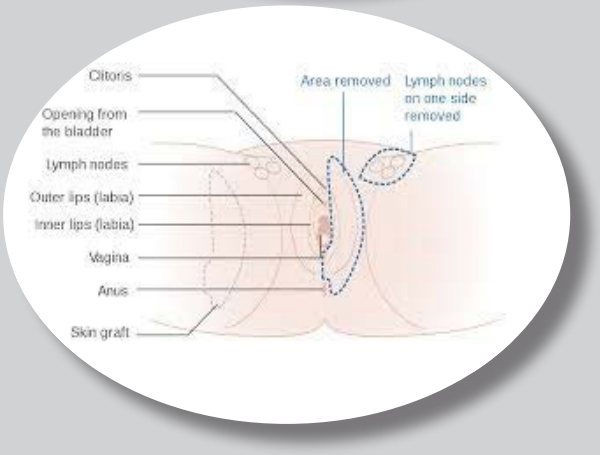


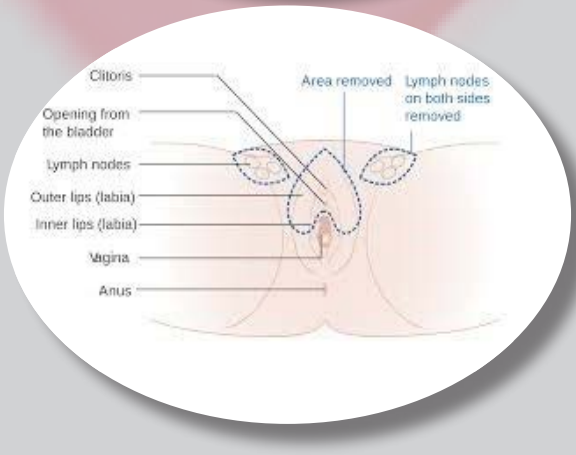
Vulvectomy

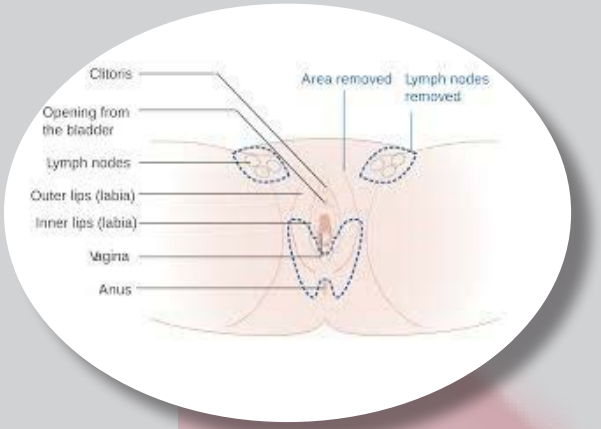
Skinning/simple vulvectomy



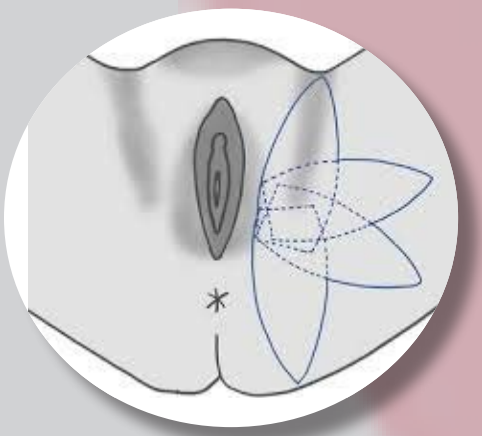








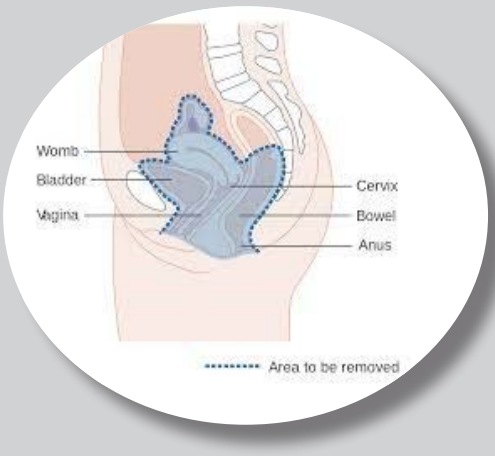
Sometimes vulvectomy may also include reconstruction of the vulva where flaps of skin are moved from the buttocks (or other sites) to help with healing and aesthetics.

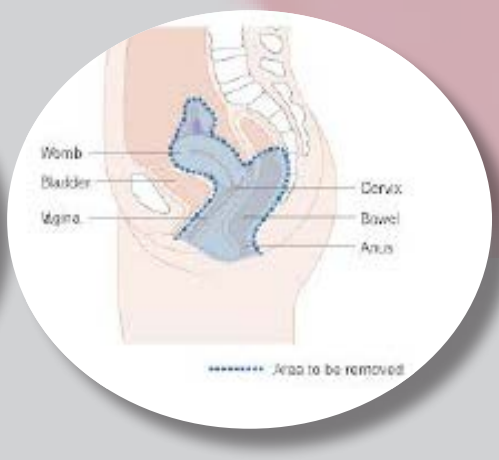


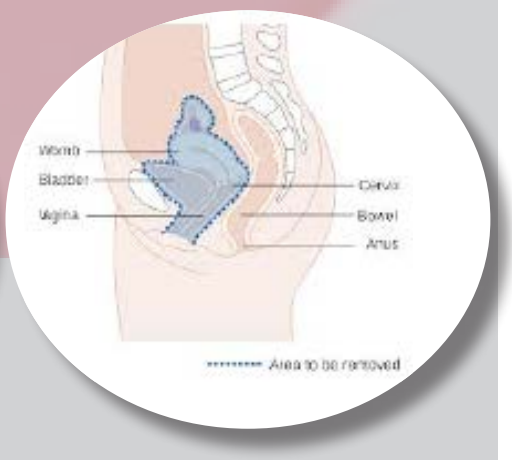
Exenteration

Exenteration is a very radical operation performed with the intention of cure. Location of the tumour will inform what tissues need removing, this could be the bladder and/or part of the bowel including the anus. Plastic reconstruction may be needed to enable the wound to be closed, sometimes the vagina is also reconstructed although this will very much depend on the patient.

This procedure will result in formation of stoma(s) – colostomy and/or ileal conduit although sometimes the bladder may be left and the urethra removed and a supra-pubic catheter inserted







‘Exenteration is a very radical operation performed with the intention of cure.’

Radiotherapy

Radiotherapy can be used in advanced vulval cancer, sometimes before surgery or instead if there are positive groin nodes. Radiotherapy uses radiation to destroy cancer cells, if used in conjunction with chemotherapy this is known as chemoradiation. The treatment involves rays of radiation to be sent to the part of the body that needs treating. However other healthy organs may be in the treatment field, this may cause both short and long term damage to organs eg. the bowel, bladder, vagina, pelvic bones and lymphatics.

Chemotherapy

Chemotherapy can be used to try and shrink cancer before surgery, with radiotherapy in nodal disease or in metastatic and recurrent disease.

Reflect on case study

- Reflect on the case study and consider what treatment Carol had. Look back at the pictures and consider possible interventions she would have needed if she came through your practice area. Consider your needs and barriers in caring for women with vulval cancer.
- Reflect on your learning through this resource, make a note on your reflection of 5 things you have learnt.

End of life

- Some women will need end of life care due to vulval cancer.
- Care should fit with meeting the holistic needs of the woman and her family.
- Interventions should be aimed at symptom control, this may include management of the vulval lesion.

Side effects

Consider the impact of each treatment, both short and long term.

Surgery:
- Changed anatomy, effect on sexual enjoyment, passing urine may have changed if surgery near urethral meatus – spraying or other problems.
- Opening bowels may also be impacted if surgery near anus.
- Lymphoedema if lymph node dissection.
- Pain/discomfort/scars.
- Sexuality issues – body image, femininity etc.

Radiotherapy
- Radiotherapy can damage the tissues it comes into contact with.
- There may be skin changes both in appearance and structure (fibrosis, less elasticity, irritated etc), bladder problems including incontinence, urgency, and recurrent UTI.
- Bowel – from constipation through to diarrhoea, incontinence, malabsorption.
- Sexual function problems.
- Lymphoedema.
- Fatigue.

Chemotherapy
- Hair loss (potentially), nausea and sickness, fatigue, anaemia, risk of infection – all whilst on treatment.
- Long term fatigue.

Partner's needs

The partners of women with vulval cancer may also need support to help them to understand the diagnosis, treatment and possible long-term effects of treatment. Involvement of partners should always be with the permission of the woman.

Within the case study John was present at every point of Carol's care and involved in all conversations regarding treatment and the implications of treatment to both Carol and their intimate relationship. Involving and supporting both of them enabled them to develop coping mechanisms and strategies to jointly overcome the challenges they were faced with.

Consider that diagnosis and treatment may impact on the couple, what strategies you would need to support them?

Further reading

- Suspected cancer: recognition and referral (2 week wait) - NICE guideline (NG12) Last up-date July 2017 https://www.nice.org.uk/guidance/ng12
- Guidelines for Diagnosis and Management of Vulval Carcinoma https://bgcs.org.uk/Vulval%20Cancer%20booklet%2020052014.pdf
- Guidance on long term consequences of treatment for gynaecological cancer: Pelvic radiotherapy. https://www.macmillan.org.uk/documents/aboutus/health_professionals/mac14942_gynae_guide.pdf
- Female sexual health after a cancer diagnosis. https://rcni.com/sites/rcn_nspace/files/cnp.14.7.16.e1206.pdf
- Information on vulval conditions http://lichensclerosus.org/ & http://www.vulvalpainsociety.org/vps/

'Radiotherapy can be used in advanced vulval cancer, sometimes before surgery or instead if there are positive groin nodes.'