

Vulvar and Vaginal Cancer

VULVAR CANCERS

Malignant tumours of the vulva are uncommon, representing about 4% of malignancies of the female genital tract. Most tumours are squamous cell carcinomas, with melanomas, adenocarcinomas, basal cell carcinomas, and sarcomas occurring much less frequently.

Squamous cell carcinoma of the vulva occurs mainly in postmenopausal women, and the mean age at diagnosis is 65 years. A history of chronic vulvar itching is common.

EPIDEMIOLOGY

Recent studies suggest two different etiologic types of vulvar cancer. One type is seen mainly in younger patients, is related to human papillomavirus (HPV) infection and smoking, and is commonly associated with vulvar intraepithelial neoplasia (VIN). The more common type is seen mainly in elderly women and is unrelated to smoking or HPV infection; concurrent VIN is uncommon. It is often associated with lichen sclerosus.

INTRAEPITHELIAL NEOPLASIA (PRECANCER)

There are two varieties of intraepithelial neoplasia: squamous cell carcinoma in situ (Bowen's disease) or VIN III, and Paget's disease.

Squamous Cell Carcinoma In Situ: VIN III

During the last 25 years, the incidence of VIN has increased. Younger patients are being affected, and the mean age is approximately 45 years.

Clinical Features

Itching is the most common symptom, although some patients present with palpable or visible abnormalities of the vulva. Most lesions are elevated, but the colour may be white, red, pink, grey, or brown. Approximately 20% of the lesions have a "wart-like" appearance, and the lesions are multicentric in about two thirds of cases.

Diagnosis

Careful inspection of the vulva in a bright light, with the aid of a magnifying glass if necessary, is the most useful technique for detecting abnormal areas. Colposcopic examination of the entire vulva after the application of 5% acetic acid will sometimes highlight additional areas.

Management

The mainstay of treatment is local superficial surgical excision. Margins of about 5mm are usually adequate. For extensive lesions, a skin graft may be required.

Laser therapy is also effective, particularly for multiple small lesions, or for lesions involving the clitoris or perianal area.

PAGET'S DISEASE

Paget's disease of the vulva predominantly affects postmenopausal white women. It is an adenocarcinoma in situ.

Itching and tenderness are common and may be long-standing. The affected area is usually eczematoid in appearance. In 10% to 20% of cases, Paget's disease is associated with an underlying adenocarcinoma.

Management

Local superficial excision with 5- to 10-mm margins is required to clear the gross lesion, exclude underlying invasive cancer, and to relieve symptoms. Recurrences are common and may be treated by further excision or laser therapy. If an underlying invasive carcinoma is present, the treatment should be the same as for other invasive vulvar cancers.

INVASIVE VULVAR CANCER

SQUAMOUS CELL CARCINOMA

Squamous cell carcinoma accounts for about 90% of vulvar cancers. Patients generally present with a vulvar lump, although long-standing itching is common. The lesions may be raised, ulcerated, pigmented, or warty in appearance, and definitive diagnosis requires biopsy under local anaesthesia.

Methods of Spread

Vulvar cancer spreads by direct extension to adjacent structures, such as the vagina, urethra, and anus; by lymphatic spread to regional lymph nodes; and rarely via the blood stream to distant sites, including the lungs, liver, and bone.

Staging

The FIGO staging system is shown below.

International Federation of Gynecology and Obstetrics (FIGO) staging of vulvar carcinoma (1994)

Stage

Stage 0	Carcinoma in situ, intraepithelial carcinoma
Stage I	Tumour confined to the vulva or perineum, or both, and 2cm or less in greatest dimension; no nodal metastasis
Stage Ia	As above with stromal invasion \leq 1mm
Stage Ib	As above with stromal invasion $>$ 1mm
Stage II nodal	Tumour confined to the vulva or perineum, or both, and more than 2cm in greatest dimension; no metastasis
Stage III	Tumour of any size with: 1. Adjacent spread to the urethra and/or vagina, and/or the anus. 2. Unilateral regional lymph node metastasis, or a combination
Stage IV	
Stage IVa	Tumour invades any of the following: upper urethra, bladder mucosa, rectal mucosa, pelvic bone or bilateral regional node metastasis, or a combination
Stage IVb	Any distant metastasis including pelvic lymph nodes

Management

EARLY VULVAR CANCER

Patients with Stage Ia disease (i.e., tumours in whom the depth of penetration is less than 1mm) do not need groin dissection. All other patients require at least removal of the groin lymph nodes on the side of the tumour. Postoperative radiation is required if there are positive lymph nodes.

The primary cancer should be treated by a wide and deep resection (radical local excision). Surgical margins should be at least 1cm.

ADVANCED VULVAR CANCER

If the cancer involves the proximal urethra, anus, or rectovaginal septum, radiation or chemoradiation to shrink the primary tumour, followed if necessary by more conservative surgical excision should be performed. Bilateral groin node dissection, or at least removal of any large, positive nodes is usually performed prior to the radiation therapy.

Prognosis

The overall survival rate for vulvar carcinoma is about 70%. Patients with negative nodes have a survival of about 90%.

MALIGNANT MELANOMA

Malignant melanoma is the second most common type of vulvar cancer. They occur predominantly in postmenopausal white women.

Diagnosis and Staging

Any pigmented lesion on the vulva requires excisional biopsy for histologic diagnosis. The prognosis correlates more closely with the depth of penetration into the dermis. Those lesions that penetrate to a depth of 1mm or less rarely metastasise.

Management

For the superficial lesions referred to previously, wide excision, with margins of at least 1cm, is adequate therapy. For lesions with ≥ 1 mm invasion, **wide excision of the primary tumour is usually combined with removal of groin lymph nodes.**

Prognosis

The overall 5-year survival rate for vulvar melanomas is approximately 30%.

VAGINAL CANCERS

INTRAEPITHELIAL NEOPLASIA (PRECANCER)

Carcinoma in situ of the vagina (VAIN) is much less common than its counterparts on the cervix or vulva. Most lesions occur in the upper third of the vagina, and the patients usually have no symptoms.

Vaginal intraepithelial neoplasia is related to infection with the human papilloma virus (HPV) in many cases. Patients with a past history of precancer or cancer of the cervix or vulva are at increased risk.

Diagnosis

The diagnosis is usually considered because of an abnormal Pap smear in a woman who either has had a hysterectomy or has no demonstrable cervical abnormality. Definitive diagnosis requires vaginal biopsy, which should be directed by colposcopy or Lugol's iodine staining of the vagina.

Management

Surgical excision is the mainstay of therapy, and this may require excision of the vaginal apex. At times, extensive disease requires total vaginectomy and creation of a new vagina using a skin graft. Laser therapy and 5-fluorouracil (a chemotherapy) are alternatives to surgical excision.

SQUAMOUS CELL CANCER OF THE VAGINA

Squamous cell carcinoma of the vagina is uncommon. The mean age of patients at presentation is about 60 years. Symptoms consist of abnormal vaginal bleeding, vaginal discharge, and urinary symptoms. Punch biopsy is required to confirm the diagnosis.

Patterns of Spread

Vaginal cancer spreads by direct invasion as well as by lymphatic and blood stream dissemination. **Direct tumour spread may result in involvement of the bladder, urethra, or rectum, or progressive lateral extension to the pelvic side wall.**

The FIGO staging for vaginal cancer is clinical, as shown below.

International Federation of Gynecology and Obstetrics (FIGO) staging of vaginal cancer

Stage	Description
Stage I	Carcinoma limited to the vaginal wall
Stage II	Carcinoma has involved the subvaginal tissue but has not extended onto the pelvic side wall
Stage III	Carcinoma has extended to the pelvic side wall
Stage IV	Carcinoma has extended beyond the true pelvis or has involved the mucosa of the bladder or rectum
Stage IVa	Spread to bladder or rectum
Stage IVb	Spread to distant organs

Management

Radiotherapy is the main method of treatment for primary vaginal cancer. Initial treatment usually consists of external irradiation to the pelvis to shrink the primary tumour and treat the pelvic lymph nodes. Brachytherapy is then given. When the lower third of the vagina is involved, the groin nodes should either be included in the treatment field or surgically removed.

Prognosis

The overall 5-year survival for vaginal cancer is about 50%.