About Vulvar Cancer

Overview and Types

If you’ve been diagnosed with vulvar cancer or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- What Is Vulvar Cancer?

Research and Statistics

See the latest estimates for new cases of vulvar cancer and deaths in the US and what research is currently being done.

- Key Statistics for Vulvar Cancer
- What’s New in Vulvar Cancer Research and Treatment?

What Is Vulvar Cancer?

The vulva is the outer part of the female genitals. The vulva includes the opening of the vagina (sometimes called the vestibule), the labia majora (outer lips), the labia minora (inner lips), and the clitoris.
Around the opening of the vagina, there are 2 sets of skin folds. The inner set, called the **labia minora**, are small and hairless. The outer set, the **labia majora**, are larger, with hair on the outer surface. (Labia is Latin for lips.) The inner and outer labia meet, protecting the vaginal opening and, just above it, the opening of the urethra (the short tube that carries urine from the bladder). The Bartholin glands are found just inside the opening of the vagina -- one on each side. These glands produce a mucus-like fluid that acts as a lubricant during sex.

At the front of the vagina, the labia minora meet to form a fold or small hood of skin called the **prepuce**. The **clitoris** is beneath the prepuce. The clitoris is an approximately ¾-inch structure of highly sensitive tissue that becomes swollen with blood during sexual stimulation. The labia minora also meet at a place just beneath the vaginal opening, at the **fourchette**. Beyond the fourchette is the **anus**, the opening to the rectum. This is where stool comes out of the body. The space between the vagina and the anus is called the **perineum**.

Cancer of the vulva (also known as vulvar cancer) most often affects the inner edges of the labia majora or the labia minora. It starts in the clitoris or in the Bartholin glands less often.
Types of vulvar cancer

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see What Is Cancer?1

Squamous cell carcinomas

Most cancers of the vulva are squamous cell carcinomas. This type of cancer starts in squamous cells, the main type of skin cells. There are several subtypes of squamous cell carcinoma:

- The keratinizing type is most common. It usually develops in older women and is not linked to infection with human papilloma virus (HPV) (HPV is discussed in Risk Factors for Vulvar Cancer2).
- Basaloid and warty types are less common. These are the kinds more often found in younger women with HPV infections.
- Verrucous carcinoma is an uncommon subtype that's important to recognize because it's slow-growing and tends to have a good prognosis (outlook). This cancer looks like a large wart and a biopsy is needed to be sure it's not a benign (non-cancer) growth.

Adenocarcinoma

Cancer that starts in gland cells is called adenocarcinoma. About 8 of every 100 vulvar cancers are adenocarcinomas. Vulvar adenocarcinomas most often start in cells of the Bartholin glands. These glands are found just inside the opening of the vagina. A Bartholin gland cancer is easily mistaken for a cyst (build-up of fluid in the gland), so it's common to take awhile to get an accurate diagnosis. Most Bartholin gland cancers are adenocarcinomas. Adenocarcinomas can also form in the sweat glands of the vulvar skin.

Paget disease of the vulva is a condition in which adenocarcinoma cells are found in the top layer of the vulvar skin. Up to 25% of patients with vulvar Paget disease also have an invasive vulvar adenocarcinoma (in a Bartholin gland or sweat gland). In the remaining patients, the cancer cells are found only in the skin's top layer and have not grown into the tissues below.

Melanoma
Melanomas are cancers that start in the pigment-producing cells that give skin color. They are much more common on sun-exposed areas of the skin, but can start in other areas, such as the vulva. Vulvar melanomas are rare, making up about 6 of every 100 vulvar cancers.

More information on this can be found in Melanoma Skin Cancer3.

Sarcoma

A sarcoma is a cancer that starts in the cells of bones, muscles, or connective tissue. Less than 2 of every 100 vulvar cancers are sarcomas. Unlike other cancers of the vulva, vulvar sarcomas can occur in females at any age, including in childhood.

Basal cell carcinoma

Basal cell carcinoma, the most common type of skin cancer, is more often found on sun-exposed areas of the skin. It occurs very rarely on the vulva. For more information on this type of cancer, see Basal and Squamous Cell Skin Cancer4.

References

See all references for Vulvar Cancer
(https://www.cancer.org/content/cancer/en/cancer/vulvar-cancer/references.html)

Last Medical Review: November 2, 2017 Last Revised: January 16, 2018

Key Statistics for Vulvar Cancer

In the United States, vulvar cancer accounts for nearly 6% of cancers of the female reproductive organs and 0.7% of all cancers in women. In the United States, women have a 1 in 333 chance of developing vulvar cancer at some point during their life.

The American Cancer Society's estimates for vulvar cancer in the United States for 2019 are:
• About 6,070 cancers of the vulva will be diagnosed
• About 1,280 women will die of this cancer.

Visit the American Cancer Society’s Cancer Statistics Center for more key statistics.

References

See all references for Vulvar Cancer
(https://www.cancer.org/content/cancer/en/cancer/vulvar-cancer/references.html)

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What's New in Vulvar Cancer Research and Treatment?

Vulvar cancer is rare, which makes it hard to study. Still, research is being done to find new ways to prevent and treat cancer of the vulva. There are some promising new developments.

Oncogenes and tumor suppressor genes

Scientists are learning more about how certain genes called oncogenes and tumor suppressor genes control cell growth and how changes in these genes cause normal vulvar cells to become cancer. This information is already being used to develop new drugs that counteract the effects of these gene changes. The ultimate goal of this research is gene therapy. Gene therapy involves replacing the damaged genes in cancer cells with normal genes in order to stop the abnormal behavior of these cells.

Drug treatments

Researchers are learning more about the gene and protein changes that take place in vulvar cancer cells. Drugs that target changes like these are already being used to treat
other cancers. There have been case reports of using these drugs known as targeted therapies\(^2\) to treat vulvar cancers, too. These drugs do not have the same kind of side effects as traditional chemo drugs do. So far, the drugs cetuximab and erlotinib have been tried and doctors have reported some success in a few patients. Sometimes cetuximab is combined with cisplatin chemotherapy for treatment. These drugs need further study.

**Surgery, radiation therapy, and chemotherapy**

Clinical trials are being done to determine the best way to use and combine surgery\(^3\), radiation therapy\(^4\), and chemotherapy\(^5\). These trials will provide information about whether certain groups of patients benefit from radiation after surgery and whether patients with cancer that has spread to lymph nodes\(^6\) benefit from chemotherapy or pelvic radiation therapy.

The use of internal radiation therapy, called brachytherapy, along with external beam radiation is being studied. This form of radiation is done by placing tiny pieces of radioactive material right into the tumor. It's already used to treat other types of cancer, and women with certain vulvar tumors might benefit from it, too. More research is needed to find out if and when this treatment might improve treatment outcomes.

**Identifying lymph nodes**

Another area of interest is lymph node mapping. Vulvar cancer can spread to lymph nodes in the groin. Better ways to look for this spread and identify nodes with cancer might help doctors treat these nodes and decrease the risk of cancer coming back there. It could also allow them to save the healthy nodes and decrease the risk of long-term swelling in the groin and legs, called lymphedema\(^7\).

**Hyperlinks**

5. https://www.cancer.org/content/cancer/en/cancer/vulvar-
References

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