

EXTRA-MAMMARY PAGET'S DISEASE



What is extra-mammary Paget's disease?

What are the symptoms?

What do I see?

Extra-mammary Paget's disease (EMP) is a rare, superficial skin precancer or cancer. It is almost exclusively a disease of postmenopausal women, usually older than 60 years. It can be primary, when its origin is in the vulva or secondary when it comes from other adjacent regions like urethra, cervix, bladder or anorectal area. Usually it grows superficially, but in up to 25% of the cases grow deeply and it presents as an invasive cancer. The area near the anus has the higher risk of having an underlying cancer.

The most common symptom is itching. The patient usually refers reports a history of vulvar pruritus for over 2 years and treatments with different creams or ointments for other skin problems. If the skin cracks, then it can feel irritated, sore, or painful. Bleeding may occur, particularly if the skin is scratched. Sometimes there are no symptoms at all.

The rash of EMP can look like eczema as it can be red, scaly and crusty. The skin surface may become soggy from moisture and white dotted areas may be seen. The rash is usually on the outer lips (labia) but can spread around the anus and to the inner surface of the vulva. It may be on one side or both sides of the vulva. It is important to see a specialist because EMP can mimic other vulvar conditions such as fungal infection, dermatitis (eczema) or psoriasis.

How is it **diagnosed**?

The diagnosis is made by biopsy. This is a simple procedure in which a small sample of the lesion is removed under local anesthesia to be analyzed by a pathologist. Some special techniques may be used in order to give more information.

What other tests **will I need**?

Patients with Paget's disease of the vulva should be evaluated for the possibility of another cancer. Twenty to thirty percent of patients with vulvar Paget's disease have another cancer (breast, ovary, urethra, bladder, rectum, or cervix). The risk depends on the site of the EMP. At times, special tests are done on the pathology specimen. For example, if EMP is present around the anal margin your doctor will usually arrange for the bowel to be screened by colonoscopy (a special test where a camera is passed in to the bowel). If EMP is present around the opening to the bladder (urethra) then your bladder may be investigated. Other tests that might be arranged but won't be necessary in all cases include urine cytology, ultrasound or CT and mammogram. Your doctor will discuss with which of these tests are needed.

Treatment of patients initially diagnosed with noninvasive Paget's disease of the vulva consists of surgical excision of the vulva with 2 cm margins. There is no need to remove the lymph nodes if there is no invasion. Deep excision is not necessary. If the disease is invasive, a deeper excision is performed along with lymph node dissection, at times.

Radiation therapy and chemotherapy in the treatment of Paget's disease of the vulva are also not well defined. At times they are used for patients who are not suitable for surgery. Topical imiquimod has also been used. It is generally reserved for recurrent Paget's disease of the vulva, but there are reports of its use as an initial treatment. Imiquimod is an immunomodulator that encourages the immune system to destroy the abnormal cells. The advantages of this treatment are that it is a self-applied cream and it preserves vulvar anatomy however it can make the skin very sore during treatment. Although it is not yet licensed for this disease, the results are promising. The doctor will discuss with the patient the best treatment options for each case. 20 to 30 percent vulvar Paget's disease will have, or will develop an adenocarcinoma at another location of the body.

Will I need **follow-up?**

Yes. It is important that you are followed-up regularly to check the response to the treatment and to detect any areas of disease that may come back (recurrences occur in 20-50% of the patients). The vulva should be inspected at least annually for recurrence with vulvoscopy.