



EXTRA-MAMMARY PAGET'S DISEASE

What is extra-mammary Paget's disease?

Extra-mammary Paget's disease (EMP) is a rare, superficial skin 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 perineum or secondary when it comes from other adjacent regions like urethra, cervix, bladder or anorectal area. Usually it grows superficially, but in approximately 20% of the cases it can grow deeply and it turns into an invasive cancer.

What are the symptoms?

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 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.

What do I SEE?

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 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 funga 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 such as immunohistochemistry in order to give more information

What other tests will I need?

In a small number of cases EMP can be associated with other cancers. The size of the risk depends on the site of the EMP and the special tests 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.

What is the treatment?

The usual is local surgical excision however recurrence is frequent. The excision should be wide enough to get a disease-free margin, usually 1.5 to 2 cm away from the visible lesion. There is no need to remove the lymph nodes if there is no invasion.

Radiotherapy – It uses superficial X-rays to destroy the tissue.

Photodynamic therapy (PDT)- It uses a tumor- localizing photosensitizer, 5-aminolevulinic acid (ALA), in combination with nonthermal light of an appropriate wavelength to generate oxygen-induced cell death. Because PDT has been shown to be very effective in the treatment of nonmelanoma skin carcinomas, recently is being proposed for the management of Paget's disease with promising results.

Medical treatments –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 pathology, the results are promising. The doctor will discuss with the patient the best treatment options for each case.

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 20-40% of the patients).