Understanding Extramammary Paget's Disease: A Guide for Patients

Zaim Haq

What is Extramammary Paget's Disease?

Extramammary Paget's Disease (EMPD) is a rare skin cancer that primarily affects areas of your body with a lot of apocrine glands. For most patients, this condition is most commonly found in the genital and anal regions of the body, and specifically the vulva in female patients. EPMD often appears as red, scaly, and itchy patches on the vulva, which can eventually become thickened over time. As a result, EMPD is often mistaken for eczema or dermatitis due to their similar appearance and location[1, 2, 3].

The apocrine glands are connected to the unusual changes seen in EMPD. These are a type of sweat gland that produces and secretes a thicker, oily sweat, mainly for cooling the body and in response to stress[4]. In EMPD, the primary problematic cells are called Paget cells. These cells can infiltrate, or spread, to adjacent structures within the skin, such as sweat ducts and hair follicles, eventually leading to EMPD. While EPMD can be a slow-growing cancer, in some cases, it may be associated with underlying cancers in nearby tissues or organs[1].

Why Do People Get EMPD?

The exact cause of EMPD is not well understood, but it is thought to be related to the abnormal growth of cells in the epidermis, the skin's outermost layer. There are two main types of EMPD: primary, where the disease originates in the skin itself, and secondary, where the disease spreads to the skin from an internal cancer. Here are a few factors that might increase the risk of developing EMPD[1, 2, 5]:

- Age, with most cases being diagnosed in individuals over the age of 50.
- Gender, with women being more likely to develop the condition than men, particularly in the vulvar location.
- Chronic irritation or inflammation in the affected area.
- A history of other skin conditions or cancers.

How Do You Know If You Have EMPD?

EMPD often presents as a persistent red, scaly patch that might itch or burn and is typically found in the anogenital areas, though it can appear in other areas as mentioned. Due to its resemblance to more common skin conditions like eczema, it can be challenging to diagnose. If your doctor suspects EMPD, they will perform a biopsy, taking a small sample of the affected skin for examination under a microscope to confirm the diagnosis[2, 5].

What Happens If You Have EMPD?

Following a diagnosis of EMPD, your physician may recommend further tests to check for any associated underlying cancers, particularly if the EMPD is of the secondary type. This might include imaging studies or additional biopsies of nearby tissues[2, 5].

Treating EMPD

The treatment for EMPD primarily involves surgical removal of the affected skin. Wide local excision is commonly used, where the lesion along with a margin of healthy skin is removed to ensure all cancerous cells are taken out. In some cases, Mohs micrographic surgery might be considered. This technique involves removing the cancerous tissue layer by layer and examining each layer under a microscope until no cancer cells are detected, helping to conserve as much healthy tissue as possible[5, 6].

Non-surgical options may include topical therapies, photodynamic therapy, or radiation, especially for patients who cannot undergo surgery or for treating large areas affected by EMPD[5,7].

Looking After Your Skin

Whether you've had EMPD or not, it's crucial to maintain good skin health. This includes regular self-examinations of your skin, particularly in areas prone to EMPD like the vulva, and reporting any persistent changes to your doctor. Additionally, maintaining a healthy lifestyle and seeking regular medical check-ups can be beneficial.

Protecting your skin and being aware of changes can help in early detection and treatment of EMPD and other skin conditions. Remember, your skin's health is a vital part of your overall well-being, and taking care of it is essential for maintaining your quality of life!

Citations

1. Yildiz P, Ronen S, Aung PP, Trinidad C, Kajoian A, Prieto VG. Extramammary Paget Disease-A Challenging Case. *Am J Dermatopathol*. 2019;41(11):867-868. doi:10.1097/DAD.0000000000001311
2. Ishizuki S, Nakamura Y. Extramammary Paget's Disease: Diagnosis, Pathogenesis, and Treatment with Focus on Recent Developments. *Curr Oncol*. 2021;28(4):2969-2986. Published 2021 Aug 5. doi:10.3390/curroncol28040260
3. Simonds RM, Segal RJ, Sharma A. Extramammary Paget's disease: a review of the literature. *Int J Dermatol*. 2019;58(8):871-879. doi:10.1111/ijd.14328
4. Farkaš R. Apocrine secretion: New insights into an old phenomenon. *Biochim Biophys Acta*. 2015;1850(9):1740-1750. doi:10.1016/j.bbagen.2015.05.003
5. Asel M, LeBoeuf NR. Extramammary Paget's Disease. *Hematol Oncol Clin North Am*. 2019;33(1):73-85. doi:10.1016/j.hoc.2018.09.003
6. Hendi A, Brodland DG, Zitelli JA. Extramammary Paget's disease: surgical treatment with Mohs micrographic surgery. *J Am Acad Dermatol*. 2004;51(5):767-773. doi:10.1016/j.jaad.2004.07.004
7. Tolia M, Tsoukalas N, Sofoudis C, et al. Primary extramammary invasive Paget's vulvar disease: what is the standard, what are the challenges and what is the future for radiotherapy?. *BMC Cancer*. 2016;16:563. Published 2016 Jul 29. doi:10.1186/s12885-016-2622-5

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Citations

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