



[Authoritative facts](#) about the skin from the [New Zealand Dermatological Society Incorporated](#).

[Home](#) | [Site age specific](#)

## Extramammary Paget disease

Extramammary Paget disease is an uncommon cancer characterised by a chronic eczema-like rash of the skin around the anogenital regions of males and females. Under the microscope it looks very similar to the more common type of [mammary Pagets disease](#) that occurs on the breast. Extramammary Paget disease most commonly occurs in women aged between 50–60 years.

About 25% of extramammary Paget disease is associated with an underlying cancer, either in situ (i.e. confined to the outside layer of the tissue of origin) or a more widespread invasive cancer. Sometimes the extramammary Pagets disease may be present for 10–15 years before evidence of cancer or metastases appear.

### What are the signs and symptoms?

The most common symptom is a mild to intense itching of a lesion found around the groin, genitalia, perineum or perianal area. Pain and bleeding may occur from scratching lesions that have been around for a long time. Thickened plaques may form that can become red, scaly and crusty. Although they may appear similar to [eczema](#), they fail to clear up with [topical steroid](#) creams.

#### Extramammary Paget disease



Vulvar Pagets disease



Axillary Pagets disease



Vulvar Pagets disease

In women the most common area involved is the vulva. First symptoms are usually itching and burning of lesions that may persist for a long time. These may spread to the labia, mons pubis, vagina and thighs. Perianal lesions may extend up into the anal canal.

The location of extramammary Paget disease is useful in predicting the risk of associated cancer. For example, it appears that 25–35% of extramammary Paget disease situated around the perianal region is associated with an underlying colorectal cancer.

### Diagnosis of extramammary Paget disease

[Skin biopsy](#) of the lesion is performed to get an accurate diagnosis of extramammary Paget disease as there are several other [genital skin diseases](#) that may appear similar. Under microscopy, the presence of Paget cells along with other histological findings confirms diagnosis. Special stains may be necessary to distinguish Paget's disease from early [melanoma](#) (melanoma in situ).

### What is the treatment of extramammary Paget disease?

Margin-controlled surgical excision of the affected area to an adequate depth is the standard treatment for extramammary Paget disease (Mohs surgery). The margin is sometimes difficult to define particularly when lesions are spread sporadically throughout the anogenital region. Recurrence is common so patients should be re-examined every 3 months after surgery for the next 2 years, after which annual follow-ups are recommended.

The immune response modifier, [imiquimod](#) cream, appears to be a promising additional therapy.

#### Related information

---

#### References:

Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

#### On DermNet NZ:

- [Genital skin diseases](#)
- [Vulvar intraepithelial carcinoma](#)
- [Penile intraepithelial carcinoma](#)
- [Skin metastasis](#)

#### Other websites:

- [Extramammary Paget Disease](#) - emedicine dermatology, the online textbook

#### Books:

See the [DermNet NZ bookstore](#)

**Author:** Vanessa Ngan, staff writer

DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

Created 2003. Last updated 26 Aug 2007. © 2008 NZDS. Disclaimer.