EXTRA-MAMMARY PAGET DISEASE

What are the aims of this leaflet?

This leaflet has been written to help you understand more about extra-mammary Paget disease. It tells you what it is, what causes it, what can be done about it, and where you can find out more about it.

What is extra-mammary Paget disease?

Extra-mammary Paget disease (EMPD) is a rare, slow-growing disease that is a pre-invasive form of skin cancer. It looks similar to a patch of eczema and usually affects skin in the genital area and around the anus. It is commonest in people aged over 50, with studies showing it peaks at around age 65. It is less common in males and in skin of colour.

Primary EMPD arises from cells in the skin, whilst secondary EMPD is due to spread of pre-cancerous or cancerous cells from nearby or distant organs/regions, most commonly the opening to the bladder (urethra), cervix, bladder or bowel.

Paget disease, in contrast, refers to the same type of changes occurring in the breast or nipple. There is no relation between EMPD and another disease called Paget disease of the bone.

What are the symptoms?

The most common symptom is itching in the affected skin. Often people have had long-term itching over years which has failed to respond to moisturisers or steroid creams. Pain and bleeding may occur, particularly if the skin is scratched. 1 in 10 patients will experience no symptoms at all.

What does extra-mammary Paget disease look like?

The rash of EMPD can look like eczema, with red, scaly and sometimes weeping patches. In skin of colour it may appear as dark, scaly, weepy patches. The skin surface may become thickened with white raised areas. There is often a clearly visible border between affected & unaffected skin. The rash is usually on the outer lips (labia) of the vulva in women and the perianal area in men. The scrotum, groin and penis are less commonly affected. It may be on one side or both sides.

How is it diagnosed?

The diagnosis is made by skin biopsy. This is a simple procedure in which a small sample of the lesion is removed under local anaesthesia and analysed under a microscope. Special laboratory staining techniques may be needed in order to give more information on the type of cells involved.

What other tests will I need?

In a small number of people EMPD is associated with other cancers, and further tests to look for these may be required. These may include blood tests, ultrasound/CT/MRI scans and in some cases biopsies of other parts of the body like lymph nodes (glands) in the groin. If EMPD is present around the anus or around the opening to the bladder (urethra), a camera test of the bowel or bladder may be arranged to investigate these areas.

What is the treatment?

The usual treatment is surgery to remove the area; however, recurrence is frequent. The surgery should be wide enough to make sure that all the EMPD is removed with clear skin, usually 1.5 to 2cm away from the visible problem. Mohs micrographic surgery is a specialised type of skin surgery that can be used to treat EMPD and has been shown to reduce recurrence rates. There is usually no need to remove the lymph nodes (glands) in the groin, unless a biopsy test shows that they are also affected.

Non-surgical treatments include imiquimod cream, which encourages the immune system to destroy the abnormal cells, laser and a form of light treatment (photodynamic therapy). Radiotherapy is an alternative treatment in elderly patients or those unfit for surgery. It may also be used in combination with surgery to treat recurrence.

If EMPD has spread from an internal organ of the body, then this will probably also require treatment.

Will I need follow-up?

Yes. It is important that you are followed-up regularly by a dermatologist to check the response to the treatment and to detect any areas of disease that may come back as recurrence is common.

Where can I get more information?

Web links to detailed leaflets:

http://www.dermnetnz.org/site-age-specific/extra-mammary-paget.html

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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