EXTRA-MAMMARY PAGET'S DISEASE

What are the aims of this leaflet?

This leaflet has been written to help you understand more about extra-mammary Paget's 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's disease?

Extra-mammary Paget's disease (EMPD) is a rare, slow-growing disease that is usually due to a pre-invasive type of skin cancer. Usually it is confined to the skin, but in approximately 20% of the cases it can be associated with an invasive cancer more deeply. It typically looks similar to a patch of eczema. It usually affects skin in the genital area and around the anus of both males and females. It is commonest in people aged between 50-60 years. It can be primary, when its origin is in the skin, or secondary, when it comes from other adjacent regions internally like urethra, cervix, bladder or bowel. Paget's disease, in contrast, refers to the same type of changes affecting the breast or nipple. There is no relation to another disease called Paget's disease of the bone.

What are the symptoms?

The most common symptom is itching in the affected skin. Often patients report a history of chronic itching over years which has failed to respond to moisturisers or topical steroid creams. Pain and bleeding may occur, particularly if the skin is scratched. Sometimes there are no symptoms at all.

What does extra-mammary Paget's disease look like?

The rash of EMPD can look like eczema, with red, flaky or sometimes weeping patches. The skin surface may become thickened with white raised areas. The rash is usually on the outer lips (labia) of the vulva in women and the penis or scrotum in men,

but it can spread backwards to the perineum and anus. It may be on one side or both sides of anogenital skin.

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 to be analysed by a pathologist. Some special 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 cases EMPD can be associated with other cancers, and further tests to look for these may be required. For example, ultrasound scans of lymph nodes, or if EMPD is present around the anus your doctor will usually arrange for bowel investigations. If EMPD is present around the opening to the bladder (urethra) then your bladder may be investigated.

What is the treatment?

The usual treatment 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 in the groin, if there is no invasion.

Other medical treatments mainly used if required for recurrences include:

Imiquimod cream, which encourages the immune system to destroy the abnormal cells. Radiotherapy, laser and a form of light treatment (photodynamic therapy) can also be used.

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

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

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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