Paget's Disease, Extramammary

http://www.aocd.org

Extramammary Paget’s disease is a malignant condition that affects skin other than the breast/nipple (mammary) area. It is named after Sir James Paget, the surgeon who first described the condition. It is an adenocarcinoma that grows very slowly and is not invasive, meaning it does not spread beyond the confines of the skin.

It is most commonly found in the genito-urinary (GU) area involving the vulva, penis, scrotum and perineum. It can also occur in the armpit or axillary regions. Extramammary Paget’s disease is usually confined to the epidermis layer of the skin but can extend into the deeper dermal layers. It looks like a rash and is often mistaken for eczema. The symptoms are usually itching or burning similar to non-malignant rashes that can also occur in the GU area. Primary Extramammary Paget’s disease is not associated with any other underlying conditions. However it can occur in the presence of other malignancies of the GU area such as the prostate, cervix, rectum or urethra. When it present along with other malignancies in this way it is referred to as Secondary Extramammary Paget’s disease. The prognosis and treatment in these situations is related to the other underlying malignant condition(s).

Primary Extramammary Paget’s disease is typically not an invasive malignancy so surgical excision is the treatment of choice. Topical chemotherapy is sometimes used to help define the boarders of the malignancy. Intravenous chemotherapy and/or radiation treatments are not usually needed. Patients generally have a very good prognosis depending on the extent of the malignancy.