# A Case of Advanced Invasive Extramammary Paget's Disease

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Background:

Extramammary Paget's Disease (EMPD) is a rare condition of apocrine gland bearing skin. The most common site is vulval in 60%. EMPD disease can be divided into primary or secondary disease. Primary disease is of cutaneous origin and is non-invasive in the majority of cases. Those that invade beyond the vulval intraepithelial contribute 1% of all vulval malignancies[1]. Secondary disease are the result of spread of an underlying adenocarcinoma such as bladder, bowel, cervix or other site.

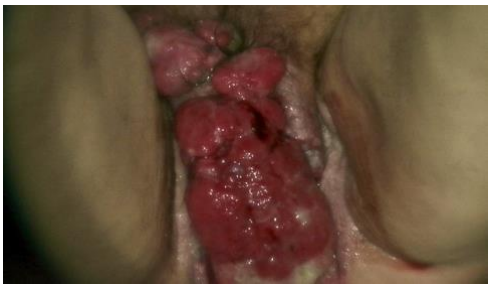
Vulval Paget's affects primarily post-menopausal caucasian women. It initially presents as an itchy lesion, often described as velveteen or eczema like. The main stay of treatment includes wide local excision, however margins are usually positive due to the diseases extension beyond the visually identified lesion, resulting in high rates of recurrence. Repeat surgeries and or use of Imiquimod and radiotherapy are commonly utilised[2]. Due to the recurrence risk, long term follow up is advised.

Case:

A 66 year old woman was admitted under gynaecology with symptomatic anaemia secondary to bleeding from her vulval lesion. Her haemoglobin was noted to be 57g/dL on admission.

The patient reported having a diagnosis of Paget's disease of the vulva made while under a private gynaecologist in 2006. At that time she had been advised to have surgical treatment which she declined due to concerns over complications and the high risk of recurrence. She was initially under 6 monthly follow up, but has not attended since 2007.

In the preceding six months the patient noted weight loss of around 5 kg. During this time she had also noted that her vulval lesion had significantly increased in size. In the last few weeks she has had intermittent episodes of heavy bleeding, soaking pads and bedding. This she believed was coming from the lesion its self. She denied any difficulty in passing urine. She had no other significant medical or surgical history.



Vulval lesion at admission

On examination the patient was found to have a large fungating vulval tumour measuring approximately 5x10cm. This likely originated from the right labia, but now obscured the whole vagina and urethra. There was also what appears to be a separate 3x4cm lesion superiorly, originating from the mons pubis on the right side. On palpation enlarged hard inguinal lymph nodes were palpable in the left. On the right either matted lymph nodes or an extension of the tumour was palpable beneath the skin.

Punch biopsy was taken and sent to histology as target. The histological appearance, together with the immunohistochemical profile, demonstrated a poorly differentiated mammary ductal-type adenocarcinoma associated with Extramammary Paget's Disease.

An MRI pelvis identified a vulval mass measuring 6.7x11cm, extending into the upper right thigh. Two additional nodules were also seen measuring 1.1cm and 2.3cm on the mons pubis and within the vagina respectively. No other adnexal masses were seen. Bone metastasis on the inferior and superior pubic rami with associated pathological fractures were seen. Right and left inguinal adenopathy, but no pelvic lymphadenopathy was identified. A CT chest, abdomen and pelvis noted pulmonary metastasis within both lung fields, with the largest lesion measuring 1.2cm.

The patient was transfused two units of red blood cells and was discharged with outpatient follow up after regional MDT. The impression at MDT was of invasive EMPD, for palliative care only. The patient sadly passed away four months later

Discussion: The 5-year survival rate is reported to be over 98% in those with noninvasive or microinvasive cases, however this is significantly reduced in those with invasive disease. One large American cohort identified a progression to invasive disease in 8% of non-invasive cases over the 25 year study period[3]. This re-enforces the need for close follow up, even in those declining treatment.

References:

1. Lam C, Finaro D. Extramammary Pagets disease: summary of current knowledge. Dermatol Clin. 2010:28(4):807-826. Available from: https://www.sciencedirect.com/science/article/abs/pii/S0733863510001270?via%3Dihub
2. Edey K, Allan E, Murdoch J, Cooper S, Bryant A. Interventions for the treatment of Paget's disease of the vulva. Cochrane database Syt Rev; June 2019; vol.6. Available from: https://doi.org/10.1002/14651858.CD009245.pub3
3. Van der Linden M, Oonk M, Van Doorn H, Bulten J, Van Dorst E, Fons G et al. Vulvar Pagets disease: A national retorsepctive cohort study. J Am Acad Dermatol 2019 Oct; 81(4):956-962. Available from: https://www.jaad.org/article/S0190-9622(18)32906-2/fulltext