

# CASE SERIES OF SCROTAL EXTRAMAMMARY PAGET'S DISEASE (EMPD) : UNCERTAINTY IN MANAGING POSITIVE MARGIN POST RECONSTRUCTIVE SURGERY

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## Introduction

Extramammary Paget's Disease (EMPD) is a rare form of malignant skin neoplasms. Originated from apocrine gland, it can be described as carcinoma in situ or can be an invasive disease. The lesions commonly develop in the vulva, penis, scrotum, perineum, perianal area, umbilicus, or axilla. There is scanty experience worldwide in terms of post reconstruction of the disease with positive surgical margins.

## Case Series

69 year old gentleman presented with 3 months history of right upper scrotal skin lesion associated with pruritis and contact bleeding. On examination, there was right inguinoscrotal erythematous plaque extending to the base of the penis. Patient underwent Wide Local Excision with split skin graft. HPE revealed Extramammary Pagets disease with microinvasion. The medial margin is involved around 5mm of microinvasion. Post operatively, the wound has been managed conservatively and 1 year later there is no evidence of recurrence over the index wound nor the distant metastasis noted.

76 year old gentlemen presented with long standing history of right scrotal skin lesion first then spread to left side associated with pruritis and pain. On examination, noted right hemiscrotum erythematous plaque extending to right thigh fold, left hemiscrotum and midshaft. Patient underwent Wide Local Excision over bilateral scrotal lesion, penile lesion with right orchidectomy with local flap. MRI staging showed extensive scrotal lesion with right inguinal nodes involvement. HPE revealed Extramammary Pagets disease with surgical margin involve over all aspect of the tissue sent. Patient underwent adjuvant radiotherapy 50Gy, 25 Fractions. No re-excision of the wound has been done and patient is disease free to date.

## Discussion and Conclusion

The standard modality of treatment in genitalia EMPD is complete resection of skin and subcutaneous tissue of the lesion. However, positive surgical margin may progress to locoregional recurrence of the disease. To date, no standard treatment for positive surgical margin.

To avoid such cases, Yang et al<sup>3</sup> suggested to have intra operative frozen biopsy analysis to avoid positive margin. However, the downside is negative intraoperative margin may have positive surgical margin in specimen and did not differ greatly compared to the standard WLE. Moh's microscopic surgery (MMS) also has been recommended as one of the manoeuvre to define the actual margins<sup>1</sup>. In view of the rarity of EMPD, the efficacy of MMS in the management of EMPD is still under investigation.

The decision of whether to re-excise or to follow up case purely defined by the prognostication of the disease<sup>2</sup>. Level or depth of tumour invasion and involvement of the nodal metastasis greatly influence the direction of surgically positive margins. While radiotherapy has a role in advanced EMPD, chemotherapy on the other hand has no clear regime to date.

Regardless of treatment manoeuvre, long-term follow-up is recommended to monitor for local disease recurrence in positive margins and to avoid progression to advanced EMPD.



Figure 1 Pre operative lesion over right upper scrotum (Case No 1)



Figure 2 Post operative with SSG (Case No 1)

### Reference

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