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Case Report

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A RARE CASE OF SCROTAL EXTRAMAMMARY PAGET'S DISEASE IN ELDERLY MALE IN DISTRICT HOSPITAL

Mohd. Shafiq Rahman*[1] and Norly Salleh[1, 2]

[1]Department of Surgery, Hospital Segamat, KM 6, Jalan Genuang, 85000 Segamat, Johor, Malaysia.
[2]Satelite CRC, Hospital Segamat, KM 6, Jalan Genuang, 85000 Segamat, Johor, Malaysia.

***Corresponding Author: Mohd. Shafiq Rahman**

Department of Surgery, Hospital Segamat, KM 6, Jalan Genuang, 85000 Segamat, Johor, Malaysia.

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ABSTRACT

Background: Extramammary Paget (EMPD) disease is a rare cancer related to Paget's disease of the breast, but found around the anus and genitals of both men and women. EPMD is rare and made up of only 6.5% cases of all cutaneous Paget's disease. ***Case Presentation:*** An 80-year-old male presented with three years history of left scrotal redness and irritation. Examination revealed widespread erythema over left scrotum and left inner thigh with two small nodules. Excision biopsy was performed. Histopathology confirmed the diagnosis of EMPD. He was subsequently referred to Plastic Surgery team for wide local excision. *Conclusions:* EMPD is rare and may take years before diagnosis is made. Awareness of this condition is important for prompt diagnosis to be made and managed accordingly.

KEYWORDS: Extramammary, Paget's Disease, scrotum, elderly.

INTRODUCTION

Extramammary Paget's disease (EMPD) was first described by Crocker in 1889, involving the scrotum and penis [1]. EMPD is uncommon. It mainly involves the perineum, perianal, scrotum and vulva. The first manifestation of the disease is usually itchiness. In most cases, diagnosis of EMPD is delayed due to similarity of presentation to other skin conditions such as dermatitis and fungal infection. Biopsy and referral to specialist center is only made after multiple, different topical treatment have failed. We report a case of scrotal EMPD in an elderly patient with pruritis and erythematous rash over the scrotal area.

CASE PRESENTATION

An 80-year-old man with background history of hypertension and major depressive disorder initially presented to our surgical clinic for left scrotal skin redness and irritation for the past three years associated with minimal serous discharge. He also complained of on and off pain, especially upon walking. He denied any weight loss or loss of appetite. He had previously used over the counter medication and ointment but no improvement noted.

Upon assessment in our surgical clinic, there was widespread erythema over the left scrotum and left inner thigh measuring 4cm x 5cm (Fig. 1). Two skin nodules noted over the left inguinoscrotal area measuring 1 cm and 2 cm each. His haemoglobin was 14.5 g/dL, white cell counts 5.5 x 10^9/L, platelet 141 x10^9/L, urea 5.6 mmol/L and creatinine 97 umol/L. The rest of his blood parameter was within normal range.

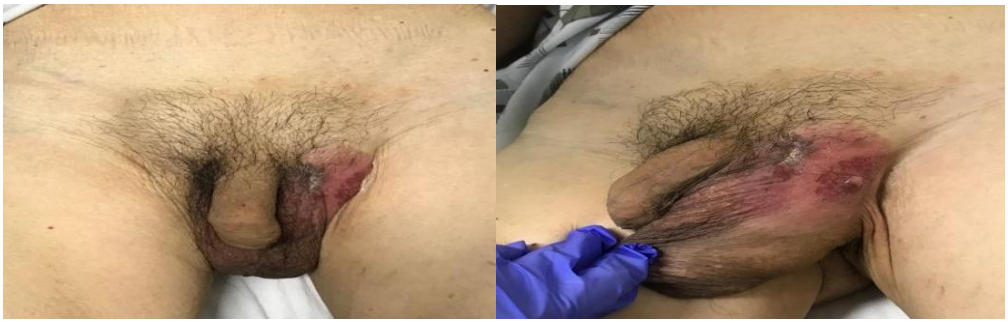

Figure 1: Left scrotal erythema with small skin nodule

He underwent excision biopsy of left scrotal wall lesion. The histopathology of the lesion confirmed the diagnosis of Extramammary Paget Disease (EMPD). However, we were unable to ascertain whether it was primary or secondary EMPD.

Contrast enhanced computer tomography (CECT) of thorax, abdomen and pelvic was performed. Prostate was enlarged measuring 3.5 cm x 4.6 cm x 3.0 cm with irregular surface indenting onto the bladder base (Figure 2). There was a well-defined rim enhancing hypoechoic lesion seen at upper inguinal region which causing bulging over the underlying skin which could be an abscess and necrotic lymph node (Figure 3). Total prostate specific antigen (tPSA) was 1.81 ng/mL which was within normal range. Patient was referred to Plastic Surgery team in tertiary hospital for wide local excision of left scrotal.

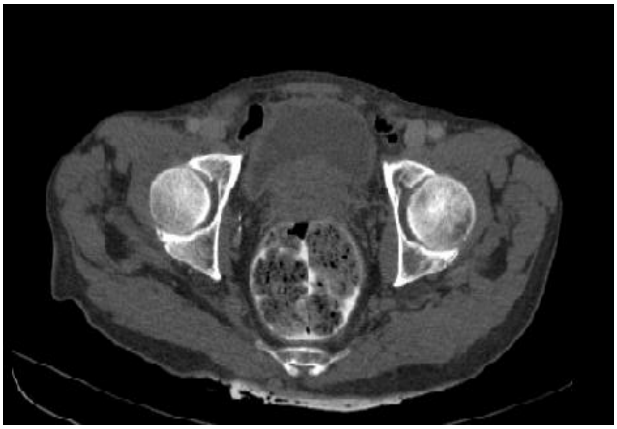

Figure 2: Prostate enlarged with irregular surface indenting onto bladder

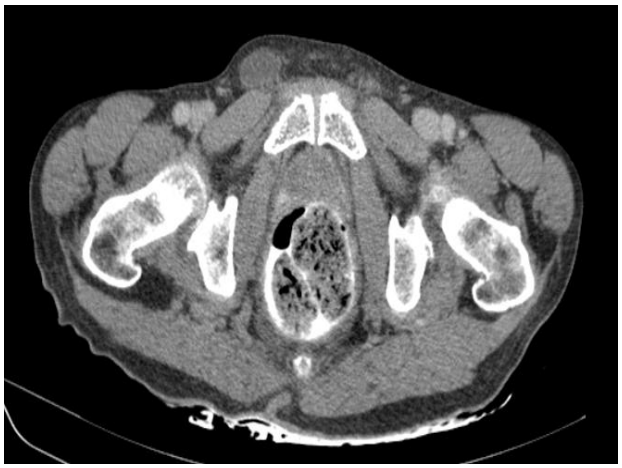

Figure 3: Right inguinal ring enhancing hypoechoic lesion.

DISCUSSION

Scrotal and penis EMPD accounts for 14% of all the EMPD cases reported, after vulva (65%) and perianal (20%) [2]. EMPD can be classified into either primary (absence of associated malignancies) or secondary (presence of associated malignancies). Primary EMPD usually involves the skin with abundant apocrine gland with no associated distant malignancies. It usually confined to epidermis, but may slowly progress and invades the dermis and spread via blood and lymphatic vessels. Secondary EMPD may be caused by epidermotropic spread of malignant cells from underlying adenocarcinoma usually involving the gastrointestinal and genitourinary tract.[2] Up to 42% of all EMPD cases has underlying malignancy, making it essential for necessary investigation and imaging to be done prior to any intervention [2]. In our case, we were unable to ascertain whether it was primary or secondary EMPD. Despite tPSA being normal, prostate biopsy may be necessary. Cystoscopy may help to ascertain whether this may be a case of secondary EMPD originating from prostate or bladder. However, due to limited resources and capability of our district hospital, this case was referred to tertiary hospital.

Diagnosis of EMPD can only be confirmed by the presence of Paget's cell, characterized by abundant pale, clear cytoplasm with enlarged pleomorphic and hyperchromatic nuclei.[3] Immunohistochemically, mucin contained within Paget's cell stained mucicarmine and periodic acid Schiff reagent. Paget's cell may also stain for cytokeratins (CK7+), carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA).[3]

Treatment of EMPD involves surgical resection with adequate margin (wide local excision), radiotherapy or systemic chemotherapy. In primary EMPD confined to epidermis, wide local excision is gold standard and the prognosis is good. Prognosis worsen once the tumour invades the dermis. Several chemotherapeutic regimes such as docetaxel monotherapy and low-dose 5-fluorouracil (5-FU)/cisplatin have been used to treat invasive primary EMPD.[4] Secondary EMPD treatment is directed toward the associated malignancy. It may involve radical surgery with or without adjuvant radiotherapy.

Regular follow up is recommended as relapses are common in all the therapeutic approaches.[1] Besides that, the associated malignancies may not be apparent at the time of diagnosis of cutaneous lesion, making it significantly important to follow up regularly and look for the early signs and symptoms of the associated malignancies.[5]

CONCLUSION

EMPD is a rare disease. Awareness of this condition is important for prompt diagnosis to be made. Failure of topical treatment with suspicious nodule should prompt the clinician to refer to respective team for tissue biopsy to avoid delay in diagnosis and treatment.

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