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A Case Series of Extramammary Paget's Disease

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Introduction

Extramammary Paget disease (EMPD) is a rare highly recurrent malignant neoplasm arising from apocrine gland-rich skin such as the vulva ,scrotum, perineum and axilla. EMPD primarily involves epidermis but may extend into the underlying dermis.It can be either a primary cutaneous adenocarcinoma or a tumour with secondary cutaneous involvement via extension from a lower gastrointestinal or urinary tract carcinoma (20-30%). Therefore, in EMPD, a synchronous tumour should always be ruled out. It's clinical features vary and is easily misdiagnosed as dermatologic diseases, hence appropriate management may be delayed. Thus,when lesions in apocrine gland-bearing regions do not heal within 1 month, the diagnosis of EMPD should be considered via biopsy. Histopathological examination of a biopsy specimen is crucial for diagnosis of EMPD.

Case reports

Case 1: 58 years old Chinese gentleman with underlying hepatitis B presented to the dermatology clinic with pruritic right scrotum lesion (Figure 1) for 3 months associated with right inguinal lymphadenopathy for the past 1 year. Biopsies were taken for both. Histopathological examination of the scrotal lesion showed EMPD and right inguinal lymph node was metastatic adenocarcinoma. Positron-emission tomography (PET) scan showed extensive nodal,bone metastasis. Patient proceeded with a wide local excision ,followed by systemic chemotherapy .

Case 2: 60 years old Chinese gentleman presented with per rectal bleeding and perianal pruritis for 1 year with no altered bowel habit or family history of malignancy. Clinical examination revealed Grade III hemorrhoids with perianal excoriation at the 3 o'clock position measuring 2.5cm diameter (Figure 2). Colonoscopy excluded presence of polyps or tumour proximally. Patient underwent Examination under anaesthesia (EUA) with stapled haemorrhoidopexy and biopsy of perianal skin lesion. Histopathology proved to be Paget's disease of the perineum(Figure 3). Patient underwent a wide local excision and reconstruction.

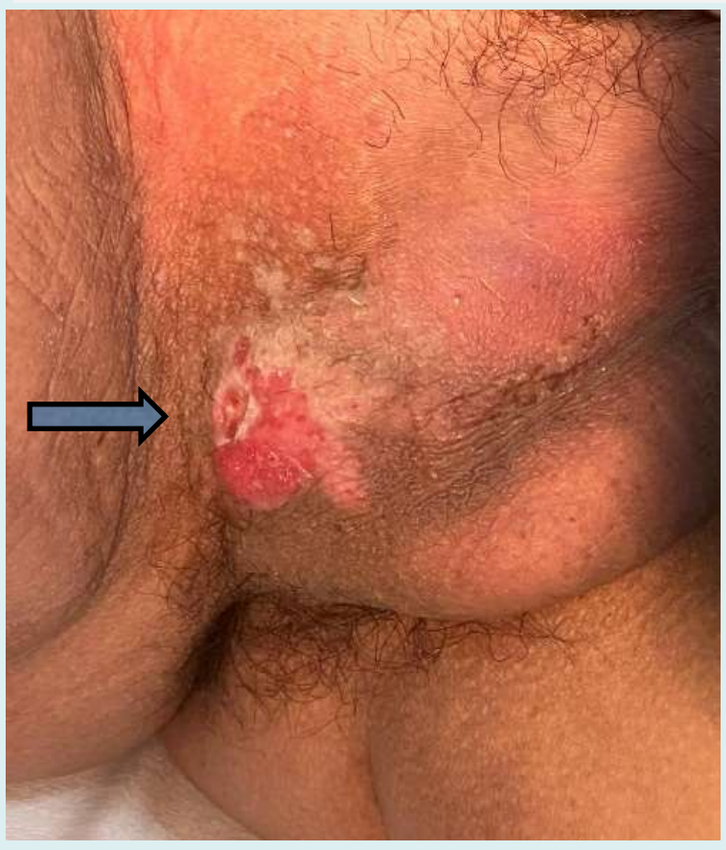

Figure 1 : 4x4cm base of right scrotum lesion

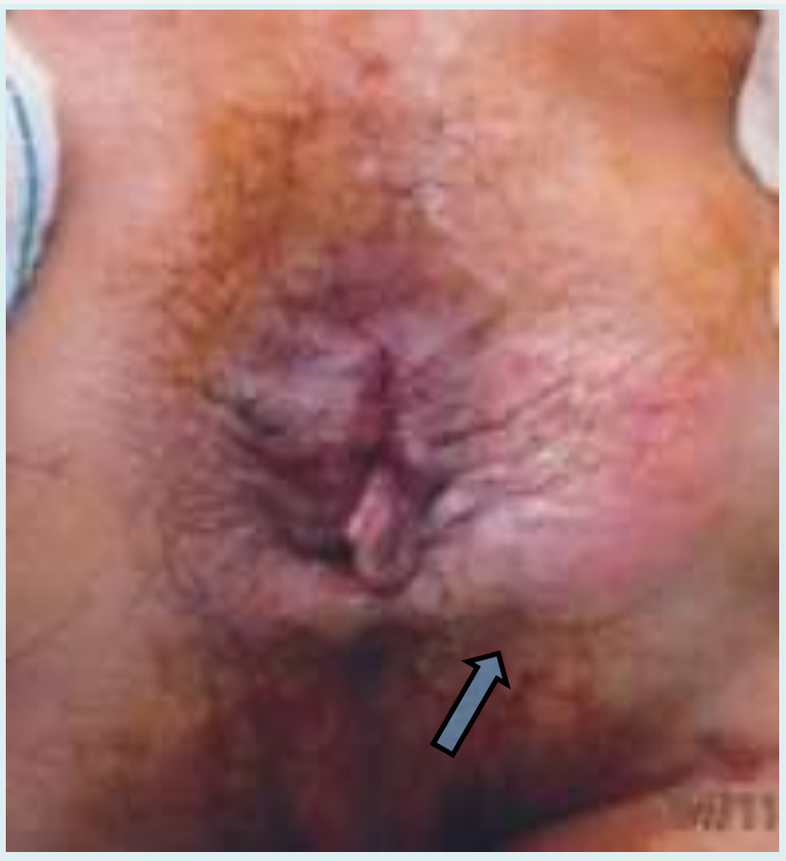

Figure 2: Perianal excoriation at 3 o clock

Figure 3 : Biopsy of the perianal lesion, showing classic pagetoid cells with pale, vacuolated cytoplasm arranged in clusters
Arrows: clustered pagetoid cells

Discussion

EMPD typically affects individuals aged after the 5th decade with four times greater prevalence in females,although a male predominance exists in Asia. The prevalence rate of EMPD is as low as 0.12 per 100 000 populations . Clinical presentation varies from being asymptomatic to burning, pruritic lesions. Typically the lesions presents as well demarcated, eczematous plaques with slightly raised edges and a red background often dotted with small ,pale islands. Differential diagnosis for EMPD includes contact dermatitis, fungal infection, eczema, Bowen's disease, and melanoma.

Generally, EMPD carries a poor prognosis, The best treatment of noninvasive EMPD is surgical resection with a 1- to 3-cm margin of grossly uninvolved tissue. As EMPD always extends beyond the clinically visible margin, Mohs micrographic surgery is preferred compared to local wide excision with a large margin.It also offers a lower rate of recurrence for EMPD (33% *vs.* 23%, respectively).However due to personal limitations, Mohs micrographic surgery could not be offered to both our patients. In patients with invasive EMPD, surgical excision is not curative. Adjuvant therapy such as radiotherapy or systemic chemotherapy is necessary. Chemoradiotherapy using 5-fluorouracil and mitomycin-C has been proven effective in inadequately excised and advanced EMPD.With early diagnosis and treatment, the five-year survival rate for patients with primary EMPD is 87%.

In our two patients, the outcome of noninvasive EMPD (Case 2) was better than in (Case 1) with advanced EMPD, although the follow-up period for Case 1 has not been long. Close follow-up is required to exclude recurrence of the disease and other concomitant malignancies,for atleast the first 5 years.

References

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