Extramammary Paget Disease: A review

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Learning points

- EMPD is a rare intraepithelial adenocarcinoma that is usually primary, but may be secondary to an underlying visceral malignancy, most commonly affecting areas of the skin which are abundant in apocrine glands.
- Diagnosis is often delayed due to the disease's quiescent nature, which can persist for years without alarming symptoms and may mimic other inflammatory or infectious dermatoses.
- In women, the vulva is the most affected area, while in men, non-anogenital skin and genital sites are similarly affected.
- Although the risk varies anatomically, EMPD cases can be linked to underlying malignancies, primarily urothelial and anorectal cancers, making screening for these essential.
- In England from 2013 to 2020, the average crude incidence rate of EMPD per year was 0.17 per 100,000 PY, with a mean of 93 cases annually and a 5-year survival rate of 74.1% in 2013.
- The primary treatment is surgical resection, either through MMS or WLE and regular follow up is recommended due to high recurrence rates.

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Abstract

Extramammary Paget disease (EMPD) is defined by the World Health Organisation (WHO), as *'a rare adenocarcinoma characterized by a predominant intraepithelial growth of neoplastic cells originating in the skin (primary EMPD) or representing intraepithelial spread of an underlying visceral carcinoma (secondary EMPD)'(1).* It affects areas rich in apocrine glands, such as genital and perianal skin. First described by Radcliffe Crocker in 1889, EMPD often remains confined to the epidermis for years, following a quiescent course. Diagnosis is frequently delayed or missed due to its insidious onset and non-specific symptoms, which can mimic other inflammatory dermatoses. Importantly, EMPD is linked to underlying visceral malignancies although the risk varies anatomically. Data from England on EMPD incidence and 5-year survival from 2013 to 2020 shows crude incidence rate of 0.17 per 100,000 person-years (PY). Additionally, the 5-year net survival for EMPD diagnosed in 2013 was 90.9%. The reported net survival for EMPD is comparable to melanoma (89.6%), prostate (85.8%) and breast cancers (85.9%) overall i.e. for all stages combined. Management of EMPD focuses on identifying and addressing any associated underlying malignancies. Other treatment options include surgery or topical treatments, while non-surgical approaches such as radiotherapy and systemic treatment are used for advanced or metastatic disease. Treatment approaches may not be curative and recurrence rates can be up to 50%, highlighting the importance of close surveillance.

This review covers EMPD's history, classification, epidemiology, aetiology, pathogenesis, diagnosis and highlights current strategies for malignancy screening and patient management. These findings are important as they contribute to our understanding of the trends and outcomes of this rare disease.

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Introduction

Extramammary Paget disease (EMPD) is an uncommon malignancy, characterised histologically and by its predilection for apocrine gland bearing areas of the skin(2).

History of EMPD

Paget's disease of the breast was first described in 1874 by Sir James Paget, who observed chronic nipple lesions that later became malignant, leading to the condition being termed 'mammary Paget's disease' (MPD)(3). In 1887, Henry Radcliffe Crocker documented the first case of extramammary Paget's disease (EMPD) in a man with a well-defined, oozing ulcerative lesion on the scrotum and penis(4). Although it initially resembled eczema, microscopic examination confirmed malignant cells(4,5). Crocker presented the case to the Dermatological Society of London(6), where Paget noted its similarity to mammary Paget's disease. EMPD has since been recognised as a rare condition affecting apocrine-rich areas and accounts for around 6.5% of all cutaneous Paget's disease cases(6).

Classification

EMPD is broadly classified into primary and secondary EMPD. Over 80% of EMPD cases are primary, emerging as a primary intraepithelial adenocarcinoma in anatomically rich areas of apocrine glands(6). When EMPD is found in non-classical cutaneous sites (without apocrine glands) such as the face, arms, back and legs, it is referred to as ectopic EMPD. Primary EMPD can further be subdivided into in-situ (confined to the epidermis), micro-invasive (<1mm of invasion), and invasive EMPD(7,8).

Secondary EMPD is less common and associated with the extension of an underlying adnexal adenocarcinoma or a visceral malignancy (range; 7-40% of EMPD cases)(9). Lesions of secondary EMPD have a similar clinical presentation as those of primary EMPD but their trajectory is of the underlying carcinoma, commonly urothelial and anorectal cancers(8). The risk and type of underlying cancer vary depending on the location of EMPD. A systematic literature review by Kibbi et al collected data from 122 studies consisting of 14,108 patients to determine association rates of internal adenocarcinoma by EMPD anatomic subtype, as shown in Table 1(10).

All patients diagnosed with EMPD should have a thorough work up to detect the presence of any underlying malignancies(6,8).

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Epidemiology

According to NHS England's National Disease Registration Service (NDRS) 'Get Data Out' (GDO) programme (11), 740 new EMPD tumours were reported between 2013-2020 in England with a mean of 93 cases per year(10). The overall crude incidence rate of EMPD per year was 0.17/100,000 person-years (PY) (95% Confidence Interval (CI) 0.14-0.20).

Epidemiological studies of EMPD using data from 22 Surveillance, Epidemiology, and End Results (SEER) Cancer Registries in the USA between 2004 and 2020 reported a crude incidence rate of 0.23 per 100,000 PY (95% CI 0.22-0.23)(11). The data also indicated a significant increase in the incidence of EMPD since 1973, with an annual rise of 3.2%(11). This increase may be attributed to the ageing population and the extended study period. Additionally, a study conducted in mainland China reported a crude prevalence of EMPD at 0.4 per 100,000 people, while in Europe the crude incidence rate was 0.7 per 100,000 PY(8).

In England, the 5-year net survival for EMPD diagnosed in 2013 was 90.9% (10). The reported net survival for EMPD was comparable to melanoma (89.6%), prostate (85.8%), and breast cancers (85.9%) overall i.e. for all stages combined(10). Additionally, the 5-year overall Kaplain-Meier (KM) (all cause) survival rate for EMPD was 74.1% (95% CI 63.4-82.1) for cases diagnosed in 2013, and 75.3% (95% CI 64.9-83.0) for those diagnosed in 2015(10). These rates are comparable to the 5-year overall survival of 73.8% (95% CI 69.8-77.3) observed across 22 SEER registries from 2004 to 2020(11). Notably, male EMPD patients in the SEER dataset had lower survival rates compared to female patients, with 5-year overall survival at 73.8% for males versus 81.3% for females, and 10-year overall survival at 53.4% for males versus 63.0% for females(11). When stratified by gender, no significant difference was found in site-specific survival rates among female patients (Chi-squared $p=0.759$), however among male patients' the overall survival was greatest for EMPD of penis and scrotum, and lowest when found in perianal and rectal regions (Chi-squared $p<0.001$)(11). Further analysis of the 10-year survival rates in England would be beneficial, along with a deeper investigation into gender-based survival differences and variations in survival based on the site of presentation.

Given that EMPD is a rare malignancy, additional studies using national cancer registries could provide additional valuable insights into its epidemiology.

Aetiology/pathogenesis

The pathogenesis of EMPD remains unclear, but in both primary and secondary forms, neoplastic cells with glandular differentiation, known as Paget cells, infiltrate the

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epidermis. These Paget cells likely originate from apocrine gland ducts or keratinocyte stem cells and spread to adjacent eccrine ducts and hair follicles, involving the full epidermal thickness. Dermal invasion is rare(6). Toker cells, which are clear epithelial cells in apocrine gland-rich areas, may act as precursors to these neoplasms, contributing to the development of primary EMPD(12).

Studies of vulvar EMPD have also shown compartmentalised T-cell infiltration with reduced intraepithelial and stromal CD4+/CD8+ cells, indicating local immune modulation(13). Downstream signalling of human epidermal growth factor receptor 2 (HER2) and involvement of the RAS/RAF-MEK-ERK and PI3K-Akt-mTOR pathways has been shown to play a key role in EMPD invasion (1). Invasion of the lymphovascular system may be due to common driver mutations in ERBB2 which encodes for HER2, as determined from a genetic analysis study of 87 EMPD lesions. Additionally, mutations in FOXA1, which is involved in breast cancer pathogenesis due to enhancing transcriptional activity of oestrogen receptor expression, is also thought to play a role in EMPD progression(14).

Clinical presentation

EMPD typically develops in older adults between the ages of 50-80 and manifests as a slow growing, ill-defined erythematous plaque which may be scaly, crusted, eroded or ulcerated (Figures 1,2)(15). Repeated trauma such as excoriations and secondary infections can lead to further morphological changes of chronic lesions and can resemble other inflammatory or infectious dermatoses such as chronic intertrigo, eczematous dermatitis or suspected tinea cruris (6). Thus, a mean delay in diagnosis of 3.8 years has been reported. Associated symptoms include pruritus, pain, oozing and sensitivity to touch from clothing, but in up to 10% of patients it can be completely asymptomatic(16).

A large analysis of 3,608 EMPD patients from 22 SEER registries (2004–2020) demonstrated sex-based differences in site involvement(11). In women, EMPD primarily affected the vulva (82.3%), then non-anogenital skin (12.8%) and the perianal/rectal region (3.8%). In men, non-anogenital skin (49.5%) and genital sites (penis/scrotum, 45.5%) were similarly common, with 4.7% involving the perianal/rectal area(11).

As part of the clinical assessment, it is important to conduct a brief systems enquiry to screen for symptoms suggestive of an underlying malignancy such as unexplained weight loss, change in appetite or bowel habits, or haematuria(6). A comprehensive physical examination should also be performed, including assessment for lymphadenopathy or

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organomegaly. Specific assessments for commonly associated malignancies are detailed in Figure 3 (17).

Diagnosis

The dermoscopic features of EMPD lesions resemble those of eczema, fungal infections, and Bowen's disease. Common features include milky-red areas, dotted vessels, glomerular vessels, polymorphous vessels, scaling and linear irregular vessels. Mun et al highlighted that milky red areas and vascular lesions were more frequently noted in EMPD lesions(18), however, Payapvipapong et al. highlighted that a "lava lake" structure of thick branching reticular white lines with intermingled white clods, and cloud-like, structureless areas may be more specific to EMPD(19). Despite these distinctions, a skin biopsy with histopathological analysis remains the gold standard for diagnosing EMPD and multiple biopsies may be required to make the diagnosis(18,19).

Histopathological findings

The hallmark of EMPD is the presence of in situ adenocarcinoma cells, sometimes in this context termed "Paget cells". These cells are large, round, or polygonal with abundant pale or vacuolated cytoplasm, prominent vesicular nuclei, and occasionally an eccentric nucleus resembling a signet ring(6). Mitoses are commonly observed. Paget cells are found either scattered or in nests within the epidermis and skin adnexal structures in continuity with the epidermis, such as hair follicles and sweat glands(6).

The epidermis typically exhibits hyperplasia, acanthosis, parakeratosis and in advanced cases, ulceration. In most cases EMPD is confined to the epidermis, however occasional cases progress with dermal invasion. When Paget cells invade the dermis, they represent an adenocarcinoma and may form gland-like structures, sometimes showing features of an apocrine carcinoma or other adnexal adenocarcinomas. Additionally, a chronic inflammatory infiltrate composed of lymphocytes, plasma cells and occasionally eosinophils is often seen in the dermis surrounding the invasive Paget cells and this reaction can mimic an inflammatory dermatosis, complicating diagnosis(6).

It is crucial to distinguish whether the EMPD is primary or secondary in origin. Immunohistochemically, primary EMPD Paget cells are strongly positive for cytokeratin (CK) 7, GATA3, and CAM5.2, and often express CEA, GCDFP15, ERBB2 (HER2), CA125. Androgen, oestrogen and progesterone receptors may be positive or negative(20). In secondary EMPD, expression of CK20 and CDX2 by the Paget cells favour a colorectal origin, uroplakin III positivity is

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typical for urothelial carcinoma, whereas expression of prostate-specific antigen (PSA) and NKX3-1 suggests an underlying prostatic adenocarcinoma(20). The abundant mucin within these cells can be identified through mucin markers, such as periodic acid-Schiff (PAS), mucicarmine, Alcian blue, and colloidal iron(20).

Due to a similar growth pattern, Paget cells in the epidermis may closely resemble melanoma in situ, Bowen's disease and T cell lymphoma, but can be distinguished by their negative reaction with immunohistochemistry to melanocytic, squamous and lymphoid markers(6,9).

These histopathological findings, combined with clinical presentation and immunohistochemical profiling, are essential for diagnosing and differentiating EMPD from other malignancies.

Staging systems and screening investigations

Limited data are available on staging in the occasional cases where there is progression of EMPD to an invasive adenocarcinoma, as it typically presents as carcinoma in situ. In a multicentre retrospective study, Ohara et al. analysed 301 patients with invasive EMPD and proposed a TNM staging system for EMPD using local tumour status, lymph node metastasis and distant metastasis as factors for classification (Table 2)(21). They also identified factors influencing survival, including tumour thickness, lymphovascular invasion, the number of lymph nodes involved, and distant metastasis(21). In this study, patients with stage IIIa (1 Lymph node metastasis) disease had a 5-year overall survival rate greater than 80% and a comparable survival curve to patients with stage II disease, indicating that early lymphatic spread might be controlled with surgical treatment(21).

Screening for underlying malignancies

The US EMPD Guideline Study Group proposed a subtype-based screening algorithm for underlying malignancy in EMPD following a systematic review of literature from 1990–2022 (Figure 3)(10). The recommended work-up varies by anatomic site and the presence of high-risk clinical or histopathologic features.

Perianal EMPD warrants comprehensive evaluation, including colonoscopy, urine cytology, and imaging of the chest, abdomen, and pelvis, due to high risk of underlying colorectal cancer. In contrast, penoscrotal EMPD and vulvar EMPD with low-risk features generally undergo more limited screening such as urine cytology, faecal occult blood testing, PSA in men, and mammography in women, reflecting the lower prevalence of associated internal malignancy.

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Cases at any site exhibiting high-risk features, including invasive histology, recurrence, multifocal disease, or immunohistochemical findings suggestive of secondary EMPD, require more detailed, organ-specific investigations tailored to the likely underlying malignancy. This risk-adapted approach aims to balance thorough detection of visceral cancers with cost-conscious, site-specific screening. However, given the rarity of EMPD and the limited universal consensus, guidelines are flexible and final evaluation should be individualised within a multidisciplinary context.

Follow-up strategies are similarly guided by expert consensus, due to scarce high-quality data. Surveillance should be risk-tailored, with closer monitoring during the first five years, selective imaging, and ongoing attention to anatomical site and disease invasiveness (22). Prospective studies and registries are needed to refine these recommendations.

Management

There are currently no EU treatment guidelines for EMPD, largely due to the lack of randomised controlled trials investigating different treatment options and small patient population owing to the low prevalence of the disease(23).

Treatment is challenging due to ill-defined clinical margins and often extensive disease at diagnosis, contributing to high recurrence rates. Options include surgical approaches such as wide local excision (WLE) and Mohs micrographic surgery (MMS), as well as non-surgical methods like radiotherapy, topical imiquimod, and systemic chemotherapy for advanced cases(24).

WLE has traditionally been the standard treatment. Due to the multifocal nature and discontinuous subclinical spread of Paget cells, a 2–5 cm margin of normal skin is typically recommended(25–27). MMS offers lower false-negative rates and tissue preservation through frozen section analysis until a negative margin is confirmed, though it is more costly and time-consuming(28). Peripheral MMS, where the tumour's edges are excised first until clear margins are achieved, followed by excision of the remaining central portion, is also an alternative suggested surgical technique(23,28).

Due to ill-defined borders extending beyond visible margins, EMPD has significant recurrence rates. Hatta et al. reported a 7.6% recurrence rate (5/66) after curative EMPD excision, unrelated to margin status(29). In contrast, Leong and Chung found recurrence rates of 15% with negative margins and 48% with positive margins(30). Long et al., analysing 154 cases, reported higher positive margins with WLE than MMS (33.3% vs. 3.4%). Positive margins increased recurrence risk 3.5-fold and were linked to worse prognosis in non-invasive disease(31).

Mapping biopsy is used to reduce positive surgical margins and the likelihood of incomplete excision(32). However, a retrospective study of 133 patients with 150 primary EMPD lesions by Kaku-Ito et al. found mapping biopsy unnecessary for well-defined lesions or when 2 cm surgical

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margins are achievable. They recommend 1 cm surgical margins for well-defined and 2 cm for ill-defined EMPD lesions(33).

If surgery is contraindicated (e.g. advanced age) or the disease is localised, alternative therapies include imiquimod 5% cream, photodynamic therapy (PDT), and radiotherapy (RT)((34). Imiquimod has been widely used in treating vulval diseases as either a neoadjuvant or adjuvant therapy following primary lesion excision. Moreover, it can be considered for primary non-surgical treatment in high-risk elderly patients (35).

Sawada et al. prospectively treated nine EMPD patients with topical imiquimod 5%, achieving a 100% response rate and 56% (5/9) achieving complete response (CR)(36). However, 60% (3/5) of CR cases recurred, likely due to untreated residual disease in skin adnexal structures(36). Snast et al. reviewed 13 studies (110 patients) and found 85% achieved ≥50% regression and 54% achieved CR, however 39% of CR patients relapsed after 29 months on average(37). Thus, imiquimod is effective but requires long-term follow-up(37).

Photodynamic therapy (PDT) is a non-invasive treatment which triggers antitumour effects but is primarily palliative rather than curative in EMPD (23,38). A review of 14 studies by Snast et al. (n=122) found a 36% CR rate and 80% showing ≥50% tumour regression, with better results using methyl-aminolevulinate (p=0.0045). However, 58% of complete responders had recurrence within 20 months, indicating limited long-term control(37).

Radiotherapy is an effective option for EMPD, particularly for patients unable to undergo surgery. Hata et al. reported a 100% initial CR in 41 patients treated with 45–80.2 Gy with 5-year local progression-free and disease-free rates of 82% and 46%, respectively(39). Dermal invasion (P=0.005) and regional lymph node metastasis (P=0.002) were associated with significantly worse survival (39). Snast et al. reinforced these findings, demonstrating a 97% CR rate with various techniques (X-ray/electron, brachytherapy, boron neutron capture), though 34% recurred after a mean 60-month follow-up(37). Recurrence rates varied by modality; highest after brachytherapy (60%) and absent after boron neutron capture therapy, though data were limited(39).

Molecular profiling has identified several actionable targets in EMPD. ERBB2 (HER2) and PIK3CA mutations occur in up to one-third of cases, implicating the PI3K/AKT/mTOR pathway in disease progression(40,41). A phase II trial reported a 76.9% response rate to trastuzumab plus docetaxel in HER2-positive metastatic EMPD, suggesting the efficacy of HER2-directed therapy(42). Metastatic tumours show oestrogen-receptor positivity in about 45% and *ERBB2* mutations independent of HER2 expression, suggesting potential benefit from both HER2-targeted and hormonal therapies(43) while androgen receptor expression correlates with tumour invasion and lymph node metastasis, supporting evaluation of androgen blockade in advanced disease(44). Transcriptomic profiling has identified dysregulation of the *FOXA1–AGR2* axis as a driver of EMPD cell differentiation, particularly in penoscrotal lesions(45), and *FOXM1* overexpression has been linked to tumour progression and poor survival, with inhibition studies demonstrating reduced cell viability(46). These findings could support the use of targeted therapies in EMPD, including HER2 or PI3K inhibitors, hormonal or androgen blockade, and emerging transcription factor–targeted therapies.

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Disease-specific survival (DSS) for EMPD is estimated at 95.9%, 92.9%, and 88.5% at 5, 10, and 15 years, respectively (34). Patients with deep dermal invasion (≥1 mm) or metastatic disease have significantly lower 5-year DSS rates of 87.1% and 52.2%, respectively, compared to those with non-invasive or microinvasive disease(47).

Five-year recurrence-free survival (RFS) was higher with MMS than WLE (63% vs. 33.7%) and superior with surgery than topical treatments (36.9% vs. 20.8%)(48). Median RFS was longest after WLE with negative margins (72 months) compared to positive margins (22 months), radiotherapy (26 months), and topical therapies (22 months)(48). Wollina et al. concluded lower recurrence (7.3% vs. 26.3%) and higher 5-year RFS (91% vs. 66%) with MMS than WLE(48,49). A recent meta-analysis reported a 7.3 % local recurrence rate after MMS vs. 26.3 % after WLE(50). Among non-surgical options, radiotherapy and imiquimod showed the most favourable 5-year RFS (30.1% and 27.6%)(47).

Given the excellent prognosis of non-invasive EMPD and the morbidity of surgery, nonsurgical options such as imiquimod and radiotherapy, with reported complete response rates of 36–54% and >50% clinical regression rates of 75–85%, are increasingly considered when MMS is unavailable or unsuitable(37,47). Furthermore, conservative management with active surveillance is considered appropriate for selected patients, especially in the context of frailty or limited life expectancy who show no evidence of invasive disease and experience minimal symptoms(51).

Conclusion

EMPD is a rare intraepithelial adenocarcinoma that primarily affects apocrine gland–bearing skin and may be primary or secondary to an underlying visceral malignancy. It often has an insidious onset and runs an indolent course, which can be associated with underlying malignancies, and occasionally progresses locally to an invasive primary adenocarcinoma. Screening for internal malignancies, surveillance and follow up strategies are largely guided by retrospective data and expert consensus at present. Diagnosis relies heavily on clinical suspicion and histopathological examination. The crude incidence and 5-year survival rates in England are similar to other published international data. However, further research using cancer registries can be used to enhance epidemiological insights into demographics, incidence, and outcomes. Surgical treatment is commonly used, although additional therapies such as PDT and radiotherapy also play important roles. Due to the frequent presence of ill-defined margins, the risk of recurrence is high and regular follow-up for these patients is recommended.

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Figure legends

Figure 1. Well-defined erythematous violaceous patch in the right groin, with evidence of oozing.

Figure 2. An ill-defined hyperkeratotic plaque with superficial ulceration more typical appearance of vulval EMPD(15).

Figure 3. Adapted algorithm showing screening for internal malignancy as recommended by Kibbi et al(17)

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Table 1. adapted from Kibbi et al showing the association rates of internal adenocarcinoma by EMPD anatomic subtype(17) .

	Perianal EMPD	Penoscrotal EMPD	Vulval EMPD	All EMPD
Rate of secondary EMPD	**28%** 79/286	**6%** 79/1310	**6%** 154/2393	**18%** 2549/14,108
Underlying adenocarcinoma (if specified)	34 colorectal	27 adnexal	24 breast	231 colorectal
	3 cervical	15 prostate	22 adnexal	83 breast
	2 prostate	10 colorectal	25 vulvar	61 prostate
	1 breast	4 renal	30 urothelial	58 urothelial
	1 bladder	2 bladder	27 endometrial	5 uterine
		2 gastric	2 colorectal	10 gastrointestinal

Table 2. Proposed EMPD stage classification. 4 EPMD stages are described, illustrated by features noted by TNM stage. Adapted from Ohara et al(21).

	0	**1**	**2**
T	Tumour in situ	Tumour thickness ≤ 4 mm **AND** no lymphovascular invasion	Tumour thickness > 4 mm **OR** lymphovascular invasion
N	No lymph node metastasis	1 lymph node metastasis	2 or more lymph node metastasis
M	No distant or lymph node metastasis beyond regional lymph node basin	Distant organ metastasis or lymph node metastasis beyond regional lymph node basin	-

	TNM Stage		
EMPD Stage Classification	T	N	M
I	1	0	0
II	2		
IIIa	Any	1	
IIIb		2	
IV		Any	1

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CPD Questions

Learning Objective: To become more familiar with the topic discussed in this article.

Question 1

Which of the following statements is TRUE regarding Extramammary Paget Disease (EMPD)?

(a) Ectopic EMPD refers to secondary EMPD found in classic sites such as the genitalia and perineum.

(b) EMPD most commonly affects the trunk and limb, with less than 15% of cases affecting the genital regions.

(c) Over 80% of EMPD cases are primary and originate as intraepithelial adenocarcinoma in apocrine gland rich areas of the skin.

(d) Primary EMPD is often associated with underlying visceral malignancies such as urothelial cancer.

(e) Secondary EMPD is more common than primary EMPD and usually arises in apocrine rich areas of the skin.

Question 2

How does Extramammary Paget Disease (EMPD) typically present in a clinical setting?

(a) A rapidly growing painful plaque usually in the groin and perianal region.

(b) A sharply demarcated hyperpigmented nodule on the upper limbs

(c) A slow growing, ill-defined erythematous plaque usually in the groin and perianal region.

(d) A well-defined painless plaque on the trunk and back.

(e) An ulcerated ill-defined violaceous plaque on the scalp.

Question 3

What is the gold standard method for diagnosis of Extramammary Paget Disease (EMPD) and subsequent classification into primary or secondary EMPD?

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(a) Clinical diagnosis alone.

(b) Dermoscopy.

(c) Histopathology.

(d) Histopathology with Immunohistochemistry.

(e) Serum tumour-marker blood tests.

Question 4

According to the proposed guidelines developed by the EMPD guideline study group in the US, what type of screening is recommended for perianal Extramammary Paget Disease (EMPD)?

(a) Comprehensive screening due to high association with internal malignancy.

(b) No screening is recommended due to low malignancy risk.

(c) Screening only if there is a family history of cancer.

(d) Screening should focus primarily on gynaecological and urological malignancies because colorectal evaluation is generally unnecessary in perianal EMPD.

(e) Targeted testing based on patient symptoms only.

Question 5

The treatment of Extramammary Paget Disease (EMPD) can be complex due to:

(a) Clinical margins are usually well-defined but surgical options are limited.

(b) It has ill-defined clinical margins which contribute to high recurrence rates.

(c) It is often diagnosed early and therefore making treatment unnecessary.

(d) The disease responding poorly to all forms of treatment.

(e) Treatment is difficult mainly because EMPD commonly metastasises before any local therapy can be attempted.

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Figure 1
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Figure 2
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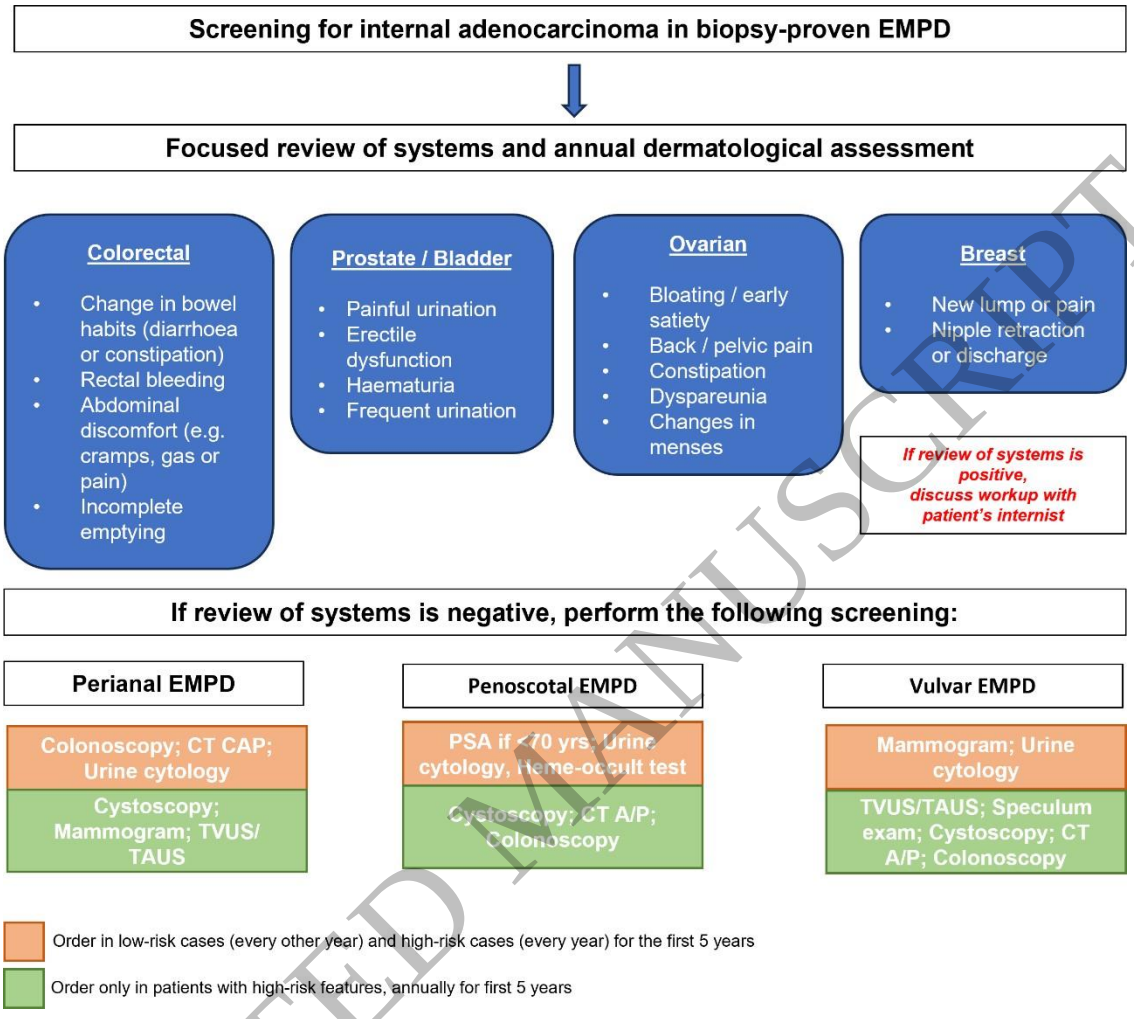


Figure 3
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