Extra Mammary Pagets Disease = اﻠﺜدﻲ ﺧارﺞ ﺑاﺠﺖ داء
Both EMPD and MPD are not preventable diseases. Rather, early diagnosis is the key to a preventive approach. Photodynamic therapy may be considered in rare cases in which surgery and radiotherapy are contraindicated.

Systemic chemotherapy has been used as an adjuvant therapy in those patients with a high risk of recurrence.

Genitourinary function due to extensive surgery that is often required for curative treatment can be compromised significantly.

Narrow surgical margins in standard excision are now being replaced by wider surgical margins. In some cases, this has resulted in a reduced rate of local recurrence.

Repeated surgeries, Mohs micrographic surgical excision (MMS) has been used to improve cure rates. Multiple scouting biopsies to help delineate the extent of the disease before surgery can be a useful adjuvant technique.

Several reviews have shown an overall recurrence rate of up to 44 percent with wide local excision. Radical vulvectomy, radical hemivulvectomy, and wide local excision have reported recurrence rates similar to those achieved with mastectomy. Proper preoperative imaging is required to rule out underlying carcinoma.

Mammary Paget disease (MPD) occurs almost exclusively in women, and the majority of cases present with a palpable mass. Given the unclear association with underlying carcinoma in EMPD, a diagnostic biopsy is ultimately performed.

Most common visceral malignancies associated with EMPD are carcinomas of the rectum, large intestine, and female genital tract. The neoplasm can then invade the dermis and lymphatics and potentially metastasize via lymphatic spread. In contrast, a smaller proportion of EMPD cases are associated with an underlying apocrine carcinoma or internal malignancy (secondary EMPD). Underlying carcinoma is not present in these cases, and these cases represent a primary intraepithelial adenocarcinoma of EMPD and MPD.

Extramammary Paget disease (EMPD) is a rare neoplasm that affects apocrine gland-bearing skin, most commonly the vulva. EMPD frequently presents as a unilateral, erythematous, scaly plaque or patch involving the vulva, often with an associated malignant lesion of the underlying mucosa. The depth of invasion appears to be an important prognostic factor, with microscopic invasive disease (less than 1 mm) having a more favorable prognosis as compared to other vulvar EMPD locations. In cases of secondary EMPD, the prognosis is worse.

Gross cystic disease fluid protein-15 (GCDFP-15) is a marker for apocrine epithelium and is frequently negative in those cases of secondary EMPD with an associated carcinoma as compared to those cases of primary intraepithelial EMPD (CK7 resistance). Immunohistochemistry is a useful adjunct in making the correct diagnosis.

Intercellular bridges are often present. These cells have a "pagetoid" appearance and simulate other intraepidermal neoplasms. The cells can be within all levels of the epidermis and can compress but preserve the basement membrane. The intraepidermal adenocarcinoma of EMPD and MPD has a similar histologic appearance.

Nipple and occasionally the areola. Ulceration and weeping with an eczematous appearance is a common presentation. The lesions can involve the areola and nipple with atypia, prominent nucleoli, and large cytoplasms. Intercellular bridges are often present. These cells have a "pagetoid" appearance and simulate other intraepidermal neoplasms.

Palpable lymph nodes are much less frequently present in EMPD. Complete physical examination is important, particularly in those patients with a palpable underlying tumor. Half have axillary adenopathy due to lymphatic spread. However, one study showed a high incidence of distant metastases in EMPD. In two cases, tumors metastasized to the lung, liver, and bone. In another case, a patient developed a brain metastasis.

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Underlying carcinoma is not present, and these cases represent a primary intraepithelial adenocarcinoma of EMPD and MPD. The malignant cells are thought to originate from intraepidermal apocrine glands or pluripotential cells of the epidermis. The neoplasm can then invade the dermis and lymphatics and potentially metastasize via lymphatic spread. In contrast, a smaller proportion of EMPD cases are associated with an underlying apocrine carcinoma or internal malignancy (secondary EMPD). Underlying carcinoma is not present, and these cases represent a primary intraepithelial adenocarcinoma of EMPD and MPD.

The differential diagnosis for EMPD and MPD includes Tinea cruris, Psoriasis, and occasionally other malignancies such as squamous cell carcinoma, melanoma, and others. Special tests may include imaging of the abdomen and pelvis, colonoscopy, barium enema, and blood work. Positron emission tomography scans may be useful for cases of invasive EMPD to evaluate for metastatic disease. Mammography is indicated in all cases of MPD, with biopsy of any underlying malignancy.

Differential Diagnosis of Mammary (MPD) and Extramammary Paget Disease (EMPD)

MPD frequently presents as a unilateral, erythematous, scaly plaque or patch involving the vulva. Most cases present with a palpable mass. In rare cases, a flat lesion may present with no palpable mass. Lesions typically involve apocrine gland-bearing skin, and the majority of cases present with a palpable mass. The lesions can involve the areola and nipple with atypia, prominent nucleoli, and large cytoplasms. Intercellular bridges are often present. These cells have a "pagetoid" appearance and simulate other intraepidermal neoplasms. The cells can be within all levels of the epidermis and can compress but preserve the basement membrane. The intraepidermal adenocarcinoma of EMPD and MPD has a similar histologic appearance.