Extramammary Pagets Disease = Extra Mammary Pagetic Disease

Extramammary Paget disease (EMPD) is a rare neoplasm that affects apocrine gland-bearing sites. It represents approximately 1 percent to 3 percent of breast malignancies. EMPD is the most common type and accounts for the majority of all patients with this disease.

Mammary Paget disease (MPD) frequently presents as a unilateral, erythematous, scaly plaque involving the breast. Approximately 15 percent of cases are associated with an underlying internal carcinoma. The most common visceral malignancies associated with EMPD are carcinomas of the rectum, stomach, and colon.

Lesions of EMPD are clinically similar to MPD and often present as a well-defined, moist, erythematous, scaly, eczematous patch. Hypo- and hyperpigmentation can occur. Burning and pruritus are common symptoms. Lesions can be accompanied by scaling, crusting, and erosions. Excessive sweating is also a frequent finding.

Histologically, EMPD can present as either intraepithelial disease or invasive carcinoma. Invasive EMPD is more likely to involve lymph nodes, especially in cases of invasive carcinoma. The clinical presentation and management of EMPD and MPD are similar. EMPD is associated with a higher risk of secondary malignancies, especially in cases of invasive carcinoma. Prognosis is generally better for EMPD than for MPD.

Diagnosis of EMPD can be challenging due to overlapping clinical and histologic features with other conditions. It is important to rule out melanoma, squamous cell carcinoma, and other apocrine tumors. Immunohistochemistry is useful in distinguishing EMPD from other conditions. The presence of MUC1 and MUC2 expression in EMPD is characteristic.

Prognosis for EMPD is generally excellent with appropriate treatment. Careful monitoring for early detection of local recurrence is critical given the potential for aggressive behavior. Surgery remains the traditional treatment for EMPD, but radiotherapy may be indicated in certain cases. PDT has been a potentially useful treatment modality in rare cases in which surgery and radiotherapy are contraindicated.

Prevention of intraepithelial disease is prudent given the limitations of current PDT technology. PDT has been shown to result in clinical and histologic cure in several case reports. This is likely due to the limited penetration of the drug and the inability to reach the deeper tumors. 5-FU may be useful as a preoperative adjunctive treatment to highlight the subclinical extent of disease before MMS or for early postoperative detection of recurrence.

Several reviews have shown an overall recurrence rate of up to 44 percent with wide local excision. The local recurrence rates are higher in cases of invasive disease as compared to non-surgical candidates and as adjuvant therapy.

Treatment of EMPD is primarily surgical. Careful monitoring for early detection of local recurrence is critical given the potential for aggressive behavior. Surgery remains the traditional treatment for EMPD, but radiotherapy may be indicated in certain cases. PDT has been a potentially useful treatment modality in rare cases in which surgery and radiotherapy are contraindicated.