Extra Mammary Pagets Disease
Both EMPD and MPD are not preventable diseases. Rather, early diagnosis is the key to prevention.

Given the patchy nature of EMPD and extension beyond clinically visible tumor, photodynamic therapy has been reported in limited cases as a potentially effective treatment. Unfortunately, photodynamic therapy is not widely available and is limited to specialized centers.

Limited reports of systemic chemotherapy for the treatment of EMPD have been described, with agents such as cisplatin, mitomycin C, epirubicin, and vincristine and docetaxel reported to have some success. However, the effectiveness of these agents is limited by the need for adequate systemic exposure and the depth of tumor invasion.

Imiquimod has been reported 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 layers of the skin where the disease is frequently found. Therefore, systemic chemotherapy is often required in addition to topical treatments.

No randomized controlled studies comparing surgery to radiotherapy have been performed to determine the superiority of one over the other. However, surgery remains the treatment of choice for EMPD when tolerated by the patient. However, high recurrence rates of up to 44 percent with wide local excision suggest that surgery is only marginally effective in preventing recurrent disease. This is likely due to the irregular margins, multi-focal nature of the condition, and subclinical extent of the disease before MMS or for early postoperative detection of recurrence.

Radiation therapy has been used as an adjuvant treatment to surgery, and limited reports of its use in EMPD have been described. However, the effectiveness of radiation therapy in EMPD is limited by the need for high doses of radiation to achieve local control, which can result in significant functional morbidity due to the anatomic location of the tumors.

Chemotherapy remains an important treatment option for EMPD, and various chemotherapy regimens have been used with varying success. The combination of cisplatin, mitomycin C, and 5-fluorouracil (5-FU) has been reported to be effective in some cases. However, the effectiveness of chemotherapy in EMPD is limited by the need for adequate systemic exposure and the depth of tumor invasion.

Overall survival in patients with EMPD is affected by lymph node status and the presence of an underlying neoplasm. Patients with negative lymph nodes have been shown to have a survival probability of 35 percent to 51 percent, as compared to 75 percent to 82 percent in patients with positive lymph nodes. Therefore, early detection and treatment of EMPD are essential to improve patient outcomes.

In cases of EMPD, workup is directed toward the possibility of an underlying gastrointestinal or genitourinary neoplasm. Imaging of the abdomen and pelvis, colonoscopy, barium enema, cystoscopy, intravenous pyelogram, chest x-ray and mammogram (for the rare association of breast malignancy) are often performed. In cases of invasive EMPD, positron emission tomography scans may be useful for cases of invasive EMPD to evaluate for evidence of distant metastasis.

Immunohistochemistry is a useful adjunct in making the correct diagnosis. It is essential to differentiate EMPD from other intraepidermal malignancies, including melanoma, pagetoid squamous cell carcinoma in situ, mycosis fungoides, and squamous cell carcinoma. EMPD is typically positive in primary EMPD not associated with underlying neoplasm. In contrast, CK20 positivity has been found more frequently in cases of secondary EMPD with underlying adenocarcinoma.

Up to one-half of all patients with MPD have a palpable underlying breast mass. Intraoperative staining with CK7 is the preferred immunostain for differentiation between primary MPD and secondary MPD. As a result, cells frequently show positive staining for periodic acid-Schiff and diastase resistant alcian blue, and positivity for MUC1, MUC2, CK20, and CD117.

Most Likely Diagnoses:
- Nummular dermatitis
- Lichen planus
- Seborrheic dermatitis
- Malignant melanoma
- Lichen sclerosus et atrophicus
- Dermatophyte infection
- Psoriasis
- Nipple eczema

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