Extra Mammary Pagets Disease = اﻠﺜدﻲ ﺧارﺞ ﺑاﺠﺖ داء
Extra Mammary Pagets Disease

Lesions of EMPD are clinically similar to MPD and often present as a well-defined, moist, erythematous, scaly plaque or patch involving the nipple and occasionally the areola. Ulceration and weeping with an eczematous appearance is frequently present. Nipple erosion and discharge may occur. Retraction of the nipple can be seen. Lesions may also occur on the vulva, perineum, and axilla. These cases are due to epidermotropic spread of malignant cells from the underlying tumor. EMPD frequently presents as a unilateral, erythematous, scaly plaque or patch involving the nipple and occasionally the areola. Ulceration and weeping with an eczematous appearance is frequently present. Nipple erosion and discharge may occur. Retraction of the nipple can be seen.

In cases of EMPD, workup is directed toward the possibility of an underlying gastrointestinal or genitourinary malignancy. Mammography is indicated in all cases of MPD, with biopsy of any underlying breast lesion to establish the diagnosis. Mucin core protein (MUC) expression is useful in the diagnosis of MPD and EMPD. GCDFP-15 is frequently negative in those cases of secondary EMPD with an associated underlying malignancy.

Laboratory tests should include markers to exclude melanoma, as both are typically negative in MPD and EMPD. Carcinoembryonic antigen (CEA) may be elevated. Cytokeratin (CK) 7 and CAM 5.2 are useful in the diagnosis of EMPD and do not react with epidermal or mucosal keratinocytes. Both Toker and Merkel cells show CK7 positivity. The cells of MPD and EMPD may stain with periodic acid-Schiff and diastase, which are strongly positive for Paget's cells. As a result, cells frequently show positive staining for periodic acid-Schiff and diastase, which are strongly positive for Paget's cells. Focal "skip areas" that are devoid of mucin, resulting in negative stains.

PATHOLOGY

CLINICAL FINDINGS

Mammary Paget disease (MPD) represents approximately 1 percent to 3 percent of breast neoplasms. The peak incidence is between 50 and 60 years of age. MPD in males is extremely rare; almost all reported cases occur in women. MPD is almost always associated with underlying in situ or invasive intraductal adenocarcinoma. Extramammary Paget disease (EMPD) is a rare neoplasm that affects apocrine gland-bearing skin of the vulva, perineum, and anus. EMPD frequently presents as a unilateral, erythematous, scaly plaque or patch involving the skin. Malignancy is present in approximately 50 percent of cases, but the majority of cases are clinically benign. EMPD is associated with underlying malignancy in a significant proportion of cases. Malignancy is present in approximately 50 percent of cases, but the majority of cases are clinically benign. EMPD is associated with underlying malignancy in a significant proportion of cases.