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Resident's Forum

RF – Screening for Internal Malignancies in Extramammary Paget Disease

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Extramammary Paget disease (EMPD) is a rare malignant epithelial neoplasm that primarily affects skin rich in apocrine glands, such as the vulvar, penoscrotal, and perianal regions. Two main forms are distinguished: primary EMPD, confined to the cutaneous and mucosal epithelia, and secondary EMPD, associated with an adjacent *in situ* or invasive internal adenocarcinoma.[1,3] Approximately 20% of cases correspond to the secondary form, with a mean interval of 5 years between the diagnosis of EMPD and detection of the internal malignancy.[2] Perianal disease carries a higher risk of malignancy than vulvar and penoscrotal EMPD.[4]

Recently, an expert panel published new recommendations for screening for internal adenocarcinoma in patients with EMPD, establishing guidelines according to anatomical location, both at diagnosis and during follow-up.[1]

All patients with EMPD should undergo the malignancy screening tests recommended for the general population according to age and sex. In addition, a review of systems is recommended at diagnosis and at every follow-up visit, with clinical suspicion-guided diagnostic testing. Moreover, a comprehensive dermatological examination is recommended annually. However, because most patients remain asymptomatic, the screening strategy should be individualized according to the anatomical location of EMPD and the presence of high-risk factors, which include:

- Bilateral inguinal erythema
- Mucosal involvement
- Dermal invasion >1 mm
- Lymphovascular invasion
- Positive immunohistochemistry for cytokeratin 20 (CK20+)

The recommendations according to anatomical location are detailed below.

Perianal EMPD

- Without high-risk factors: colonoscopy, urine cytology, and computed tomography (CT) of the abdomen and pelvis every 2 years for 5 years.
- In the presence of high-risk factors: colonoscopy, urine cytology, and CT of the chest, abdomen, and pelvis annually for 5 years. Cystoscopy is recommended in all cases and, in women, mammography and transvaginal or transabdominal ultrasound.

Vulvar EMPD

- Without high-risk factors: mammography and urine cytology every 2 years for 5 years. Some experts suggest performing cystoscopy, although no consensus has been reached.
- In the presence of high-risk factors: annual mammography and urine cytology for 5 years, with the possibility of including transvaginal or transabdominal ultrasound, speculum examination, cystoscopy, colonoscopy, and CT of the abdomen and pelvis.

Penoscrotal EMPD

- Without high-risk factors: prostate-specific antigen (PSA) testing in patients younger than 70 years, urine cytology, and fecal occult blood testing every 2 years for 5 years. Routine CT is not recommended.
- In the presence of high-risk factors: annual PSA testing, urine cytology, and fecal occult blood testing for 5 years. Annual colonoscopy, cystoscopy, and CT of the abdomen and pelvis may also be considered during this period.

In conclusion, individualized screening is recommended according to the anatomical location of EMPD and the presence of high-risk factors. In particular, a more comprehensive evaluation is advised in cases with perianal involvement due to its strong association with secondary malignancy. In contrast, vulvar and penoscrotal EMPD in the absence of high-risk factors carries a low probability of an associated malignancy. Indeed, former studies have found no increased risk in noninvasive vulvar EMPD compared with the general population.[5] Therefore, although clinical practice guidelines suggest a less intensive screening approach for low-risk vulvar and penoscrotal EMPD, the additional benefit over routine age-based screening remains unclear in these patients. In all cases, follow-up should be tailored to the level of risk and maintained for

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at least 5 years, regardless of anatomical location or initial treatment, to ensure early detection of associated malignancies.

Conflict of interest

The authors declare no conflicts of interest.

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