

Extramammary paget's disease: a real challenge for geriatricians.

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Accepted on May 17, 2017

Extramammary Paget's Disease (EMPD) is a rare intraepithelial adenocarcinoma. It mostly affects women in their seventies. EMPD develops principally in the apocrine genital, anal, and axillary zones [1]. We conducted a retrospective study at the University Hospital of Reims over a period of 20 years (1994-2014). 9 patients were included of which 7 were female. The median age of onset was 78 years (60-91). The diagnosis time ranged from a few months to 5 years prior to diagnosis. Vulvar localization remains by far the most common localization. 6 patients, all females, had pruritus (vulvar); 2 (22%) felt pain from the lesions. Looking for lymphadenopathy was positive for 3 patients (33%) with predominant localization to the inguinal area. No patient had visceral metastases. The histological makeup is the presence of cytoplasmic vacuoles of mucinous secretion. Surgery was the technique most used to treat patients, in the initial phase, as in the case of recurrences. The surgery was used in 8 patients in our study and 16 times in total. The CO₂ laser was used for 3 patients; the same applies for the medical treatment (Imiquimod). Radiotherapy was only used for a single patient. Finally, we note, for 3 patients, the combined use of 2 therapies. No patient was treated by photodynamic therapy technique. Finally, in our study, 5 patients showed a recurrence of EMPD, the majority (4/5) of whom had more than one recurrence. Thus, 3 patients had up to 3 recurrences. The average time of the occurrence of a recurrence of EMPD in our study is estimated to be 2.6 years. No deaths have been identified in our study. EMPD usually appears in the form of an erythematous patch evolving in a chronic manner, pretty well-defined, more or less infiltrated, with whitish areas, often ulcerated in the vulvar areas [2]. Pruritus is the most frequent complaint followed by a burning sensation. Surgery remains the treatment of choice, requiring modulation as a result of the comorbidities in the elderly and risk of mutilation. For standard surgery, a wide excision with a lateral margin of 2 cm is normally proposed.

Mohs micrographic surgery reduces the recurrence rate from 33% to 23% [3]. Radiation therapy is an alternative to surgery in case of the patient's refusal or contraindications to surgery, with very little toxicity. Other techniques are being studied (photodynamic therapy, Imiquimod), but their use remains limited. Searching for associated neoplasia must be systematic. Adjacent organs, preferentially affected are: colon, rectum, anus, prostate, urinary tract, female genital tract and skin. Breast cancers are also frequently found. Immunohistochemistry can serve as a prognostic marker [4]. The EGFR HER-2/neu is expressed by 30% of the MPEM cells. A p53 overexpression would predict a dermal invasion whereas the MUC5AC expression would be of good prognosis by signing the structure of the basal membrane. The CK20 expression combined with the absence of GCDFP15 (Gross Cystic Disease Fluid Protein 15) expression would then support an underlying neoplasm of the colorectal or transitional cell carcinoma type, but this hypothesis is not clear. The key is the extended monitoring of elderly subjects.

References

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