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Vulvar Merkel cell carcinoma combined with squamous cell carcinoma of the vulva

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We present a case of a 67-year-old woman who was admitted to the department of gynecological oncology with a complaint of persistent and mild pain in the area of the left labia minora. The patient has been treated with antibiotics and anti-inflammatory drugs with no improvement. She also had a dermatological consultation; however, it did not reveal dermatological causes of the patient's complaints nor any other abnormal skin lesions. A suggestion of a persistent Bartholin's gland abscess has been made. Hence, she has been referred to the surgical removal of the lesion. The general condition of the patient was stable. She reported no additional gynecologic, gastrointestinal symptoms, rapid weight loss or any other complaints.

Physical examination revealed a palpable nodular lesion about 3 cm large in the area of the left labia minora. A mild bloody discharge in the vagina was also detected. Inguinal as well as other palpable lymph nodes were not enlarged. There were no significant abnormalities on the skin of the vulva or in the transvaginal ultrasound. The abdominal ultrasound has shown no abnormalities. The patient was qualified for surgical removal of the lesion.

The oval, solid, smooth-surface tumor approximately 4 cm large has been removed during the surgery. The recovery period was uneventful, and the patient was discharged after two days. A definitive histopathological diagnosis revealed neuroendocrine neoplasm (NEN) combined with squamous cell carcinoma (SCC). Immunohistochemically, cells of NEN expressed characteristic immunophenotype: synaptophysin(+), CK7(+), CK20(+) CD56(+) whereas the squamous part was p63 (+) (Fig. 1). The microscopic morphology of NEN corresponded to Merkel cell carcinoma (MCC). Based on immunohistochemistry, primary MCC combined with SCC (MCC/SCC) of the vulva was signed. Positive surgical margins were detected. The patient has been referred to adjuvant radio-chemotherapy. Unfortunately, the patient did not return for follow-up.

NENs are rare tumors arising mainly in pancreas, lungs, and gastrointestinal tract [1]. They are rarely encountered in the gynecologic tract and constitute < 2% of female reproductive tumors. NEN of the vulva accounts for < 1% of cases. Commonly, they are admixed with other histological types, most frequently SCC [2]. Currently, WHO has divided NENs to well-differentiated neuroendocrine tumours and poorly differentiated neuroendocrine carcinomas (NECs). NECs subdivide to small-cell NECs, large-cell NECs and carcinomas admixed with NEC. This WHO classification is uniformal for NENs at majority locations, including female genital tract. However, current division has not comprised MCC [3, 4].

MCC is an aggressive, primary cutaneous neuroendocrine carcinoma, characterized by coexpression of neuroendocrine markers and CK20 (a discriminant from other types of visceral NENs) [5]. Most commonly MCC develops in elderly patients in the sun-exposed skin areas, where its occurrence is usually related to Merkel cell polyomavirus (MCV) [3, 5]. MCCs, with or without SCC component, are extremely rare in the gynecological area [1, 5].

Vulvar MCC coexistence with SCC might indicate the role of HPV in the pathogenesis of this neoplasm. The previous study determined both malignant components of the vulva incorporated the same high-risk HPV-DNA in their genome, without MCV genome. The lack of the TP53 mutation, which is related to ultraviolet light damage, was also detected.

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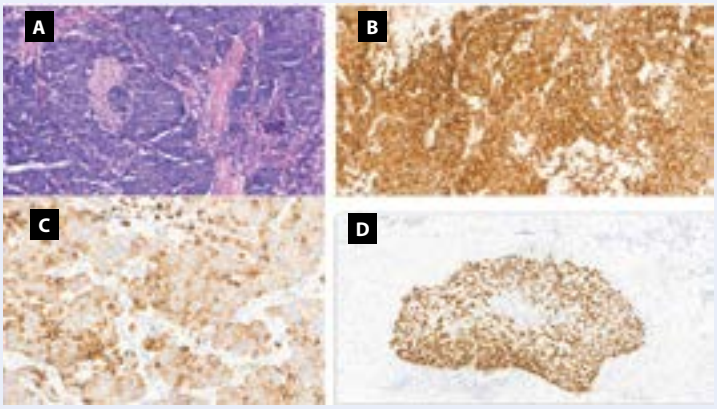

Figure 1. Vulvar Merkel cell carcinoma combined with squamous cell carcinoma. **A.** Tumor histopathology with neural invasion, hematoxylin & eosin stain, 29.56×; **B.** Neuroendocrine differentiation — diffuse synatophysin expression, immunohistochemistry stain (IS), 30.24×; **C.** Cytokeratin 20, IS, 45.79×; **D.** p63 expression in a focus of squamous cell carcinoma, IS, 26.33×

These findings support the suspicion that a vulvar MCC is HPV-related, whereas in the other origin it is more akin to be MCV-related [5].

The diagnostic process requires the assays for a possible primary lesion, as well as secondary metastases. In the presented case, based on dermatological consultation, imaging and clinical examinations, no primary lesion in the other location has been diagnosed. There are no specific guidelines regarding treatment. In clinical practices surgical excision plays a pivotal role in early-stage cases followed by adjuvant radiation or chemoradiation. Some patients might benefit from immunotherapy as well [1, 2].

We want to draw attention to the possibility of MCC (with or without SCC component) of the vulva, the importance of proper approach to the differential diagnosis and further follow-up of the patient in order to diagnose secondary metastasis. Moreover, we want to highlight the value of prophylactic vaccination against oncogenic HPV, not only to prevent anogenital SCC, but also HPV-associated vulvar MCC.

Article information and declarations

Ethics statement

Written informed consent for publication was obtained from the patient.

Author contributions

The authors confirm sole responsibility for the following: Marlena Cwynar: study conception and design; Ewa Chmielik: analysis and interpretation of results; Grzegorz Cwynar: data collection; Piotr Ptak: data collection; Karolina Kowalczyk: manuscript preparation.

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Conflict of interest

There are no conflicts of interest.

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Supplementary material

None.

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