Vulvar Merkel Cell Carcinoma – Case Report

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Abstract
Merkel cell carcinoma is a rare neuroendocrine tumour. Predisposing factors for its development are age, immunosuppression, ultraviolet light exposure, and exposure to the polyoma virus. It is usually localized in the head region, neck, and extremities. Primary neuroendocrine tumours of the vulva are extremely rare and so far there have been only 20 cases reported in the English language literature. Tumors at this location have a more aggressive clinical course than neuroendocrine tumours at other sites. The majority of patients present with metastases in regional lymph nodes at the time of diagnosis. Due to the low incidence there is no algorithm for diagnostic and therapeutic procedures.
We present a case of a 79-year-old patient with a primary vulvar neuroendocrine tumor with metastases in the regional lymph nodes. Diagnostic and therapeutic measures are described.

Key words: merkel cell carcinoma; neuroendocrine tumor; surgical management; vulva
1. Introduction

Merkel cell carcinoma (MCC) is a very rare tumor of neuroendocrine origin [1]. MCC is mostly found in the head and neck region and extremities. A primary MCC of the vulva is extremely rare and has a more aggressive course than neuroendocrine tumors of other locations [2-7]. Due to the low incidence there is no diagnostic or therapeutic algorithm [2]. We present a case of a primary vulvar neuroendocrine tumor with regional lymph node metastases in a 79-year-old patient.

2. Case report

A 79-year-old woman with recurrent hematuria was admitted to our clinic. Laboratory evaluations disclosed mild microcytic anemia and hyperlipoproteinemia. Cystoscopy did not reveal any cystic or urethral pathology. Gynaecological examination revealed a polypoid bleeding mass in the left major labia (Figure 1). A biopsy was performed and showed histological tumor fragments composed of irregularly and oval shaped fields and aggregates of interconnected tissue with atypical, small, polymorphic cells with hypercromatic nuclei and numerous mitosis but little cytoplasm. There was little connective stromal tissue between cell aggregates. Immunohistochemically tumour cells were negative for CK20, p63, HMB-45, CD20, CD3, chromogranin A, poorly focally positive for synaptophysin and diffusely positive for neuron specific enolase (NSE). Histological findings confirmed vulvar neuroendocrine carcinoma. On the next examination, the Collins test was negative and vulvoscopy revealed normal findings as well as normal cytological smears of external genital area. Preoperative X-rays of the thorax did not show any pathological processes. Transvaginal and transabdominal ultrasound detected a 1,2 cm lymph node in the left inguvinum (Figure 2).
Since more detailed tumour analysis was not possible, magnetic resonance imaging (MRI) of pelvic region was done and confirmed an unclearly demarcated tumor mass measuring 2.5 cm taking more of the left vulvar side. Both serum chromogranin A and NSE were within the normal limits. The tumor was unclearly surrounded by nearby fat tissue, did not contact the anal wall, and was separated from urethra by only a thin layer. The case was presented to endocrinology and oncology experts and the decision to perform a radical vulvectomy was made. The patient also received adjuvant radiotherapy. Despite therapy, the patient died six months after surgery.

3. Discussion

Normal Merkel cells are widely distributed in the epidermis near the end of nerve axons and may function as mechanoreceptors or chemoreceptors [3]. MCC is a very rare neuroendocrine tumour first described by Toker in 1972 [3]. Average age to develop MCC is between 70 and 90 years, and very rarely in patients under 50 years. Predisposing factors for MCC development include advanced age, immunosuppression, UV exposure, and exposure to polyoma virus [2-7]. The tumour targets the head, neck, and extremities. Primary vulvar neuroendocrine tumours are extremely rare and very few cases have been described [2,4]. It’s more aggressive than neuroendocrine tumours of other localizations and by the first examination most patients have metastases in regional lymph nodes [2,4].

MCC can only be diagnosed by histological analysis of biopsy specimens, whereas CT can identify affected surrounding tissue and lymph nodes. In addition, positive immunohistochemical staining for CK20, chromogranin A, synaptophysin, and NSE is important for definite diagnosis [3,7]. Due to the rarity of primary vulvar MCC, insufficient data is available and there are no standardized treatment protocols; therefore, all treatment guidelines are compared to extravulvar neuroendocrine tumours [2,7,10]. Radical vulvectomy including regional lymphadenectomy with positive sentinel lymph node removal is the treatment of choice [4]. Adjuvant radiotherapy is recommendable after surgery and shows better efficacy for recurrences. Survival rate depends on the tumour size and expansion [2,3,7]. Most authors recommend radical vulvectomy [8,9]. Considering the positive lymph node, survival prognosis was not promising. MCC of the vulva is universally metastatic, both to the regional lymph nodes and distant sites, and it invariably follows a rapidly fatal course. Most patients with vulvar MCC die within the first 2 years after diagnosis [10].

4. Conclusion

MCC of the vulva seems to have a more aggressive behavior than other neuroendocrine tumours and a worse prognosis than at the other sites. Early stage biopsy is essential for proper treatment and better prognosis.

Authors contribution

All authors have equally participated in data acquisition, patient follow-up, drafting the manuscript and gave their final approval.
References


