

## Case Report

## An Unusual Tumor of the Vagina

## Abstract

A 28-year-old female came with complaints of swelling in the perineal area; examination revealed a nontender and fixed mass of approximately 4 cm × 3 cm on the right vulvovaginal junction extending into the vaginal wall. Wide excision of the mass was performed; histopathological examination revealed features suggestive of Merkel cell carcinoma of vagina.

**Keywords:** *Cytokeratin, Merkel cell carcinoma, vulvovaginal mass*

## Introduction

The common tumors in the vulvovaginal junction are squamous cell carcinoma, adenocarcinoma, melanoma, sarcoma, and basal cell carcinoma. In young patients, benign lesions are mostly seen, while carcinomas are more common in older age group. Vulval carcinomas are commonly secondary to human papillomavirus-related vulvar intraepithelial neoplasia. We present a very rare tumor of the vulvovaginal junction in a young patient.

## Case Report

A 28-year-old female who was P2 L2 came to our Gynae clinic with complaints of swelling in the perineal area. On examination, a nontender mass of approximately 4 cm × 3 cm was present on the right vulvovaginal junction extending into the lower part of vaginal wall. Wide excision was performed, the histopathological examination report showed the presence of tumor in which tumor cells were arranged in the form of nests and trabeculae [Figure 1]. The individual tumor cells had small round nuclei and stippled chromatin with high nuclear: Cytoplasmic ratio with scant or moderate cytoplasm [Figure 2]. The tumor cells showed mitotic activity including atypical mitosis and perinuclear dot-like positivity with Cytokeratin. They were also positive for synaptophysin and neuron-specific enolase (NSE) indicating neuroendocrine tumors [Figure 3a-c], but

negative for MUC-1 and S-100 which are markers for squamous cell carcinoma and malignant melanoma, respectively. The final histological diagnosis was given as Merkel cell carcinoma of the vagina extending to the margins of excision. Metastatic workup was then carried out. MRI pelvis showed ill-defined heterogeneously enhancing lesion in the lower third of right vagina extending to right labia majora; there were no other metastases. She was therefore posted for repeat wide excision; there was no residual disease in the repeat specimen. She is well on follow-up at 3 months with no evidence of disease.

## Discussion

Merkel cells are components of the basal layer of the epidermis and follicular epithelium. They form clusters in areas of sensory perception, close to primary nerve endings.<sup>[1]</sup> The exact origin of the Merkel cells in the skin is unclear. The two proposed hypotheses are that they are either derived from cells in the epidermis with neuroendocrine differentiation or from the neural crest. In the human fetus, Merkel cells are first identified within the epidermis rather than the dermis. Merkel cells are not unique to skin. They are also present in the basal cell layers of normal squamous mucosa of the oral cavity in humans and other mammals.<sup>[2]</sup> In a study of the innervation of the human vagina, Hilliges *et al.* described cells that morphologically resemble Merkel cells in the basal layer of the vaginal mucosa primarily near the introitus.<sup>[3]</sup> The cells stain positive for protein gene product 9.5, a general neuronal marker, and NSE.

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**Archana,  
Saritha  
Shamsunder,  
Amit Yadav<sup>1</sup>,  
Sunita Malik**

Departments of Gynaecology  
and <sup>1</sup>Pathology, Vardhmaan  
Mahaveer Medical College and  
Safdarjung Hospital, New Delhi,  
India

## Address for correspondence:

Dr. Saritha Shamsunder,  
E 99, Ansari Nagar (East),  
AIIMS Campus,  
New Delhi - 110 029, India.  
E-mail: shamsundersaritha@  
gmail.com

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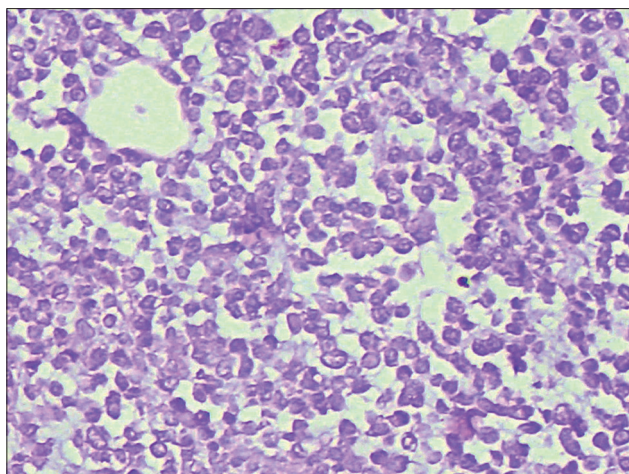


Figure 1: Tumor cells arranged in the form of nests and trabeculae (H and E,  $\times 200$ )

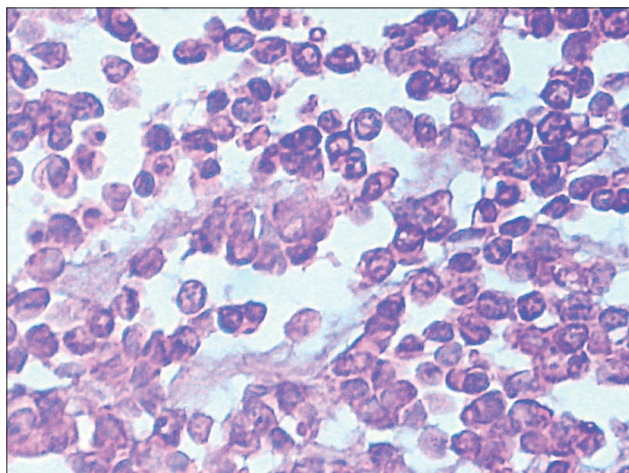


Figure 2: Tumor cells with round nuclei with stippled chromatin and increased N: C ratio (H and E,  $\times 400$ )

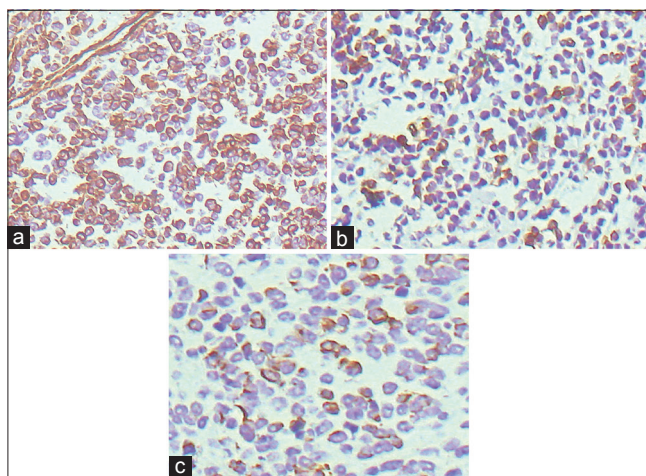


Figure 3: (a) Tumor cells showing positivity for synaptophysin. (b) Tumor cells showing positivity for neuron-specific enolase. (c) Tumor cells showing positivity for cytokeratin

Merkel cell carcinoma is a very uncommon cutaneous tumor. It is more common in head and neck and upper

extremities and typically affects elderly patients in their sixth and seventh decades.<sup>[4]</sup> Ultraviolet radiation may be the main factor responsible for the development of tumors, but viral etiology has also been debated.

Merkel cell carcinoma rarely involves the female genital system. Only 18 cases of Merkel cell carcinoma of vulvovaginal area have been reported in the literature. As these tumors are extremely rare, extensive clinical workup of the patient and characterization of the tumor are necessary to rule out metastatic disease from a different primary site. The role of CK20 is very important in this regard. CK20, a low molecular weight cytokeratin, is found in a variety of normal tissues, including intestinal epithelium, gastric foveolar epithelium, urothelium, and Merkel cells.<sup>[5]</sup> Coexpression of CK20 and NSE has been shown to reliably distinguish Merkel cell carcinoma from other neuroendocrine tumors,<sup>[6-8]</sup>

There are limited data regarding the aggressive behavior and poor prognosis of this tumor with survival rates ranging from 31% at 3 years to 74% at 5 years.<sup>[9]</sup> Merkel cell carcinoma of the vulva seems to have a more aggressive behavior and poorer prognosis than other sites.<sup>[10]</sup>

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

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