Malignant rhabdoid tumor of the vulva: A case report and review of the literature

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A R T I C L E   I N F O

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Dear Editor,

Malignant rhabdoid tumor was originally described in the kidney as a variant of Wilms' tumor in 1978 by Beckwith and Palmer [1]. Extrarenal forms of this tumor are very rare but have been described in a variety of sites including soft tissues, paraspinal and paratesticular areas, the central nervous system, colon, tongue, prostate, liver, skin, thymus, heart, pelvis, uterus, and vulva. Only 13 vulvar cases have been reported thus far in the English literature. We are reporting the 14th case and a review of the clinical features, management and prognosis of all cases.

A 51-year-old G4P3, postmenopausal woman presented to our Obstetrics and Gynecology Clinic, Erciyes University Faculty of Medicine, Kayseri, Turkey with a painless vulvar mass measuring 3 cm that was situated on the left labium major adjacent to the anterior commissura. The lesion first appeared 2 months previously and was locally excised by a gynecologist at a secondary care center with the presumed diagnosis of lipoma. The histopathological examination revealed an extrarenal rhabdoid tumor (ERT). At the time of referral to our center, 20 days after the excision, a recurrent tumor measuring 4 cm, which was painless, with irregular border and arising from the excision site was detected (Figure 1). On transvaginal ultrasonography, the uterus, endometrial thickness, and both ovaries were normal. A positron emission tomography/computer tomography with 18-F-fluorodeoxyglucose (18-F-FDG PET/CT) was performed for metastases screening. The 18-F-FDG PET/CT detected a soft-tissue lesion showing hypermetabolic activity at the left labium major, measuring 39 mm × 37 mm with irregular borders. The 18-F-FDG PET/CT image showed no lesion suspicious for metastasis. A wide local excision was performed with 1 cm surgical margin and pathologic examination confirmed ERT with negative margins.

Microscopically, tumor cells make a row of diffuse layers and prominent pleomorphism and atypia are remarkable. The tumor was composed of large polygonal cells, with extended basophilic cytoplasm. Nuclei were vesicular with distinctive nucleolus. Immunohistochemically, the neoplastic cells were diffusely positive for vimentin, CD 99, and Fli-1 and focally positive for pankeratin and epithelial membrane antigen. However, neoplastic cells were negative for cytokeratin 5/6, P63, human melanoma black monoclonal antibody 45, CD 34, desmin, S-100, actin, melan A, myoglobin, Wilms tumor-1, neuron-specific enolase, and synaptophysin, which was consistent with the diagnosis of high-grade malignant rhabdoid tumor (Figure 2).

On complete healing of the operative wound, she was referred to the Radiation Oncology Department, Erciyes University Faculty of Medicine for adjuvant radiotherapy in view of aggressive histology, grade, and size (> 3 cm). She was treated with external beam radiotherapy of 60 Gy over a period of 4 weeks (5 d/wk) to prevent local recurrence. At 12 months from her initial diagnosis, she is alive without evidence of recurrent or metastatic disease.

Vulvar carcinoma is a very rare neoplasm that accounts for about 4% of all gynecological cancers and typically affects women in the 6th decade and 7th decade of their lives. The vast majority of vulvar cancers (~90%) are squamous cell carcinoma. Other cell types that are found less frequently are malignant melanoma, anaplastic carcinomas, adenoscarcinoma of the Bartholin glands, and sarcomas [2].

ERT of the vulva is a rare but very aggressive neoplasm. Only 13 cases have been reported thus far in the English literature and six of those reported to date, died within a year of diagnosis (Table 1). The majority of reported cases of ERTs of the vulva had a history of rapidly growing, painless mass on the labium major (8 of 13 cases: 3 right, 5 left). Other reported sites are the vulva (2 cases: 1 right, 1 left), clitoris (1 case), Mons pubis (1 case), and one case at the Mons pubis adjacent to the labium major as in our case. In

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accuracy of this process is unclear because of the negative histology. Were treated with local excision plus lymph node dissection, the local excision, and 1 case unknown. Although most of the cases reported cases (9 cases with lymph node dissection, 4 cases combined surgery plus adjuvant chemotherapy [13]. The reported median survival for patients with ERT is relatively short. Six of the 13 patients treated as ERT of the vulva died within a year of diagnosis. One of the Perrone et al’s [8] patients died 138 months after diagnosis. Given the paucity of data, accurate estimation of survival in these patients is difficult, and larger series need to be reported.

In summary, ERT of the vulva is a very rare and aggressive tumor. Wide local excision of the primary and recurrent tumor is the treatment of choice. Adjuvant radiotherapy and chemotherapy do not appear to be effective in treating local and metastatic tumor. To determine a standard and effective treatment for ERT, we need larger series to accumulate data. Therefore, clinicians are recommended to present their individual cases for the establishment of guidelines for the treatment of ERT.
Conflicts of interest

The authors have no conflict of interest relevant to this article. This study was not supported by any person or institution.

References