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Case report

Unusual Complication of Ewing Sarcoma of Vulva: Case Report

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HIGHLIGHTS

- Primary Ewing sarcoma of the vulva is an extremely rare tumor.
- Urinary retention in this case is an unusual manifestation of the disease.
- Early management with multimodal therapy could help these patients.

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ABSTRACT

Introduction
Primary Ewing sarcoma of the vulva is an extremely rare tumor with less than 10 molecular confirmed reported cases. Urinary retention in this case is an unusual manifestation.

Case presentation
We describe a 15 years-old female patient with advanced Ewing sarcoma of the vulva and urinary retention during chemotherapy. The technique of catheterization with two catheters could be interesting, in this case as well. When it comes to Immunohistochemistry, tumor cells displayed CD99, NIKX2.

Conclusions
In all cells and proliferative capacity of KI-67 in 40% of tumor cells, but other markers were negative.

Keywords: Sarcoma; Ewing; Vulva Neoplasms; Vulva; Urinary Retention

Introduction

Primary Ewing sarcoma (ES) and primitive neuroectodermal tumor (PNET) are two members of the Ewing sarcoma family of tumors. These tumors are characterized by a t (11;22) (q24; q12) chromosomal translocation. Although this malignant round cell tumor is aggressive, fortunately, it is sensitive to chemotherapy. Generally, ES/PNET is unusual in the female external genital. Immunohistochemical stains and molecular cytogenetic analysis requires to confirm. Primary ES of the vulva is as an extremely rare tumor with only 10 molecular confirmed reported cases up to now. (1-3) Urinary retention in this case is an unusual manifestation in which the technique of catheterization with two catheters could be interesting, as well. Herein, we describe a 15 years-old female patient with advanced ES of the vulva and urinary retention during chemotherapy.

Case Presentation

The patient was a 15 years-old girl referred by slowly progressing swelling of the vulva from two months ago. The patient agreed to report her case after signing the informed consent and report is based on CARE guidelines. When its color changed to black the patient came to paediatrician. The patient is admitted to the hospital and underwent Magnetic resonance imaging (MRI) and tumor incisional biopsy. The patient had no history of specific clinical diseases or previous surgery. Examinations revealed vulva swelling preferably on the left side and swelling was noted over her vulvar region

measuring 15 cm × 15 cm with 7 cm × 7 cm necrosis region on the inferior left lateral site (Figure 1).

In laboratory data, creatinine was normal and urine culture was negative. The liver function test and complete blood count haven't any abnormality. Pelvic MRI showed 15 cm × 15 cm. mass in pelvis, vulva region, and spinal skeletal metastasis (Figure 2). There were multiple lung metastases on the chest x-ray, as well. The pathology report showed a malignant round cell tumor with monomorphic cells with cortical pubic bone destruction. Immunohistochemistry (IHC) study showed tumor cells displayed CD99, NIKX2 in all cells and proliferative capacity of KI-67 in 40% of tumor cells, but negative staining with desmin, myogenin, CD56, CK, EMA, synaptophysin, chromogranin, and leukocyte common antigen.

Thus ES/PNET has been concluded. In Bone marrow aspiration and biopsy, a small round cell tumor was observed, as well.

Then, the patient underwent chemotherapy with a VAC regimen (Vincristine, doxorubicin, and cyclophosphamide). Urinary retention occurred during one of chemotherapy courses. Ultrasound examination showed 600 cc of urine in the bladder. Urologic consult is done. Since the anatomy of the pubis and vulva is distorted, the meatus couldn't find, so the blind catheterization was done using a 14 Fr foley catheter. Unfortunately, this catheter came into the vagina. So, without removing this one, the second 14 Fr foley catheter inserted above the first catheter and it was successfully guided into the urinary bladder (Figure 2).

The patient was discharged with the good general condition. After four monthsduring the first months of the COVID-19 pandemic, the patient came to the emergency ward with respiratory distress and low serum oxygen saturation level. She was admitted and underwent a lung computed tomographic (CT) scan, in which a bronchopneumonia pattern was reported. COVID-19 was considered for a primary diagnosis, however, she, unfortunately, expired before COVID-19 polymerase chain reaction test.

Discussion

Primary ES of the vulva is a rare tumor with nearly 20 documented cases, and less than 10 molecular confirmed cases (4). The present patient is the eleventh molecular confirmed case in the world. She was a young female patient with advanced ES and skeletal, lung, and bone marrow metastasis. When it comes to IHC, the tumor cells in our case displayed CD99, NIKX2, and KI-67, but other markers were negative. Similar to our case, the majority

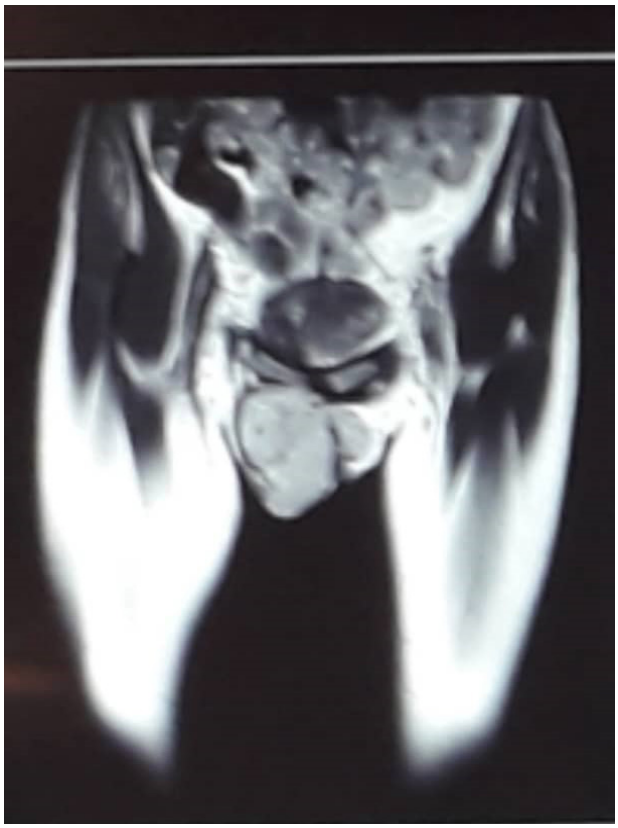

Figure 1. Magnetic resonance imaging (MRI) of the tumor

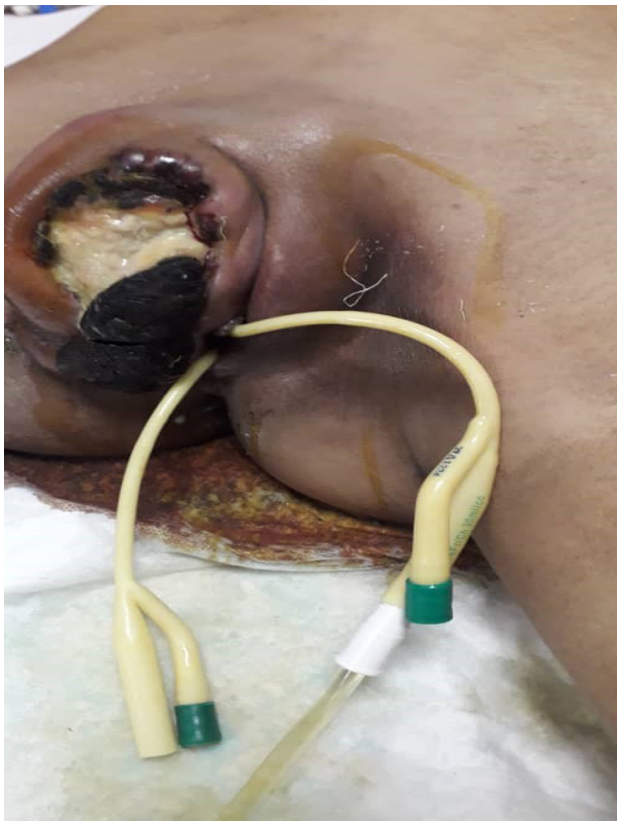

Figure 2. External appearance of the tumor after catheterization

of vulvar ES patients has CD99 stain, then LFI-1 and vimentin were the most prevalent positive stains (4). But these two markers were negative in our IHC report.

Recently, Houshemi et al reported a case of vulvar Ewing sarcoma. They have also reviewed the last three decade's similar cases. There were 20 cases, which half of them were IHC confirmed. The most prevalent site of metastasis was the lungs, then lymph nodes. However, none of the previous patients had skeletal or bone marrow metastasis. So this metastatic pattern is unique in our patient. The tumor size in the previous cases ranges from 1 to 20 cm with an average of 6.3 cm, which was smaller than our case (4). Additionally, the average age of occurrence is 22.8 years in these tumors (1), older than our patient.

The prognostic factors of ES include metastasis site (worst prognosis for bone, followed by pulmonary, then lymph node metastases) (5) tumor size (≥10 cm) (6), good histological response to neoadjuvant chemotherapy, negative margins and a multi-modal treatment by chemoradiotherapy and surgery (4). The reported longest disease-free survival (DFS) was 37 months in metastatic vulvar ES. However, there were 6 reported cases whom died of disease during or before treatment (4). Our case had three negative prognostic factors: tumor size larger than 10 cm, pulmonary metastasis, and poor response to chemotherapy. Although the survival of our patient was around 8 months, she was dead due to pneumonia.

The treatment of Ewing sarcoma requires multimodal therapy which includes surgical resection, chemotherapy, and radiotherapy. The standard chemotherapy regimen includes vincristine, doxorubicin, and cyclophosphamide, similar to our patient. However, ifosfamide and etoposide are used as an alternative in some cases. Radiation therapy is usually performed in R1/R2 cases after resection.(7) Since our patient, referred with an advanced tumor with pubic bone involvement and distant metastasis, she didn't a candidate for surgery as the first step. Thus multimodal therapy was not selected in our patient.

Conclusions

Vulvar ES is an extremely rare tumor, however, it should be considered one of the differentials diagnoses in females with vulvar lesions. Urinary retention is an unusual manifestation of this disease. IHC examination could help pathologist to find out the diagnosis. Early management with multimodal therapy could help these patients.

Authors' contributions

All authors had an equal contribution.

Ethical statements

This research has been done under the Tehran University of medical sciences committee. The patient signed the informed consent and reporting the case was based on CARE guidelines.

Funding

There is no founding.

Conflicts of interest

All authors declare that there is not any kind of conflict of interest.

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There is no acknowledgement.

Data availability

Data will be provided by the corresponding author on request.

Abbreviations

CT	Computed tomographic
DFS	Disease-free survival
ES	Ewing sarcoma
IHC	Immunohistochemistry
MRI	Magnetic resonance imaging
PNET	Primitive neuroectodermal tumor
VAC	Vincristine, doxorubicin, and cyclophosphamide

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