Case Report

Primary Ewing Sarcoma Presenting as a Vulvar Mass in an Adolescent: Case Report and Review of Literature

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A B S T R A C T

Background: Extraosseous Ewing sarcoma (ES) tumors presenting in the genitourinary tract are highly uncommon. Few cases of primary vulvar and vaginal cases of ES have been published.
Case: A 15-year-old adolescent presented with a bothersome 5-cm mass located on her left labium minorum. Following excision, a diagnosis of a primary ES was made. The patient was treated with multiagent chemotherapy and was doing well 20 months after treatment completion.
Summary and Conclusion: Based on the few available case reports and our reported case, it appears that extraosseous ES arising in superficial sites such as the vulva have better prognosis and should be treated with complete excision and multiagent chemotherapy.
Key Words: Extraosseous Ewing sarcoma, Vulvar mass, Neuroectodermal tumor, PNET

Introduction

Ewing sarcoma (ES) and primitive neuroectodermal tumor (PNET) are a group of tumors that generally affect the skeletal system.[1] Extraosseous ES and PNET presenting in the genitourinary tract are highly uncommon. Few cases of primary vulvar and vaginal cases of ES have been published to date.

We present a case of a primary ES in the labium minorum of an adolescent.

Case

A 15-year-old virginal adolescent presented with a painful mass in the left labia minora. The mass had rapidly enlarged during the past three months. She denied recent labial or vaginal trauma, fever, or vaginal discharge. Physical examination revealed a 5-cm, oblong, firm mass with smooth, well-defined contours located on the left labium minorum (Figure 1). The mass had an area of ulceration from a prior failed attempt of incision and drainage at an outside hospital. No lymphadenopathy was noted in the inguinal area. Ultrasound was performed to characterize the contents of the mass. A solid mass was noted with a moderate vascular flow. The patient was taken to the operating room for an excisional procedure. In the operating room, the area was anesthetized with 1% lidocaine, and the vulvar skin was opened around the base of the mass. The stalk was dissected away, and the base was clamped and transected. Hemostasis was obtained with a 2-0 polyglactin 910 suture, and the skin was closed with 3-0 polyglactin 910 interrupted sutures. Estimated blood loss was minimal.

The excised mass was solid in consistency, measuring 5.7 × 1.5 × 1.0 cm. The pathological evaluation revealed a malignant neoplasm with a relatively uniform appearance. The cells were round with round nuclei, coarse chromatin, inconspicuous nucleoli, and eosinophilic or clear cytoplasm (Figure 2). Immunohistochemical staining demonstrated that the cells were positive for CD99 in a membranous pattern and negative for terminal deoxyribonucleotidyl transferase (TdT), myogenin, and desmin. This combination was suggestive of an ES/PNET. Fluorescence in situ hybridization analysis for *EWSR1* gene rearrangement was negative. The reverse transcription–polymerase chain reaction (RT-PCR) analysis demonstrated the presence of an *EWSR1/ERG* fusion transcript that commonly results from the t(21;22) translocation, confirming the diagnosis of ES. The tumor was grade 2 with 5% necrosis; the specimen margins were positive for malignant cells.

The patient was referred for treatment to a pediatric oncologist. Positron emission tomography–computed tomography (PET/CT) and magnetic resonance imaging (MRI) studies were negative for metastasis. A bone marrow biopsy was performed that was negative for ES. A plan was made for 14 cycles of chemotherapy with compressed VDC/IE (vincristine, doxorubicin, cyclophosphamide/ifosfamide and etoposide). Also, a plan was made for six cycles of neoadjuvant therapy and then local control with surgery due to positive margins. Brachytherapy was planned if repeat surgical margins were positive, followed by additional cycles of chemotherapy. Partial radical vulvectomy

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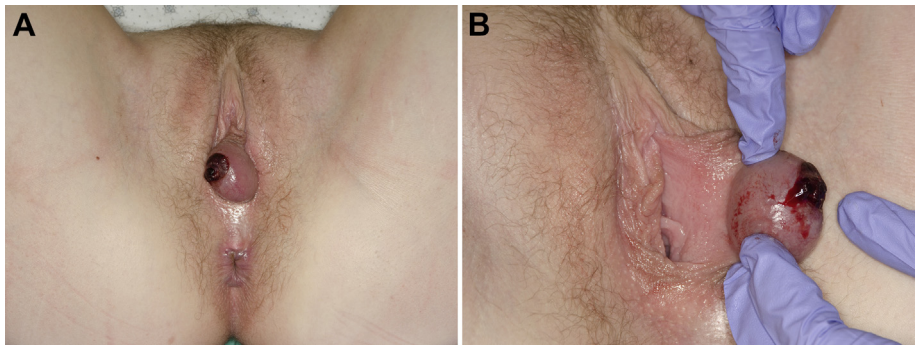

Figure 1. A 5-cm, ulcerated mass on the left labium minorum.

was performed with 1- to 1.5-cm margins around her prior excision site. The vulva was closed in three layers with interrupted 2-0 polyglactin 910 for the deep tissue and subcutaneous tissues followed by a 4-0 polyglactin 910 subcuticular stitch. Pathology revealed no residual disease.

Consequently, a more extensive groin exploration was not pursued. Because her reexcision surgery showed no evidence of disease, radiation therapy was not performed, and she subsequently completed the last eight cycles of chemotherapy. These treatments were completed nine and a half months from the initial diagnosis. Her chemotherapy therapy resulted in admissions for neutropenia, nausea, weight loss requiring a nasogastric tube for feeding, migraines, rectal fissures, and amenorrhea. These side effects resolved after chemotherapy ended, and the patient's normal menstrual function spontaneously returned.

Post treatment pelvic MRI and CT scans showed no evidence of residual disease. The patient has been followed regularly, and she remains without evidence of disease 20 months after completing chemotherapy.

Summary and Conclusion

ES typically arises in the long bones of the extremities and the bones of the pelvis.[1] Extraosseous ES uncommonly presents in the soft tissues and is exceedingly rare in the genital tract.[2]

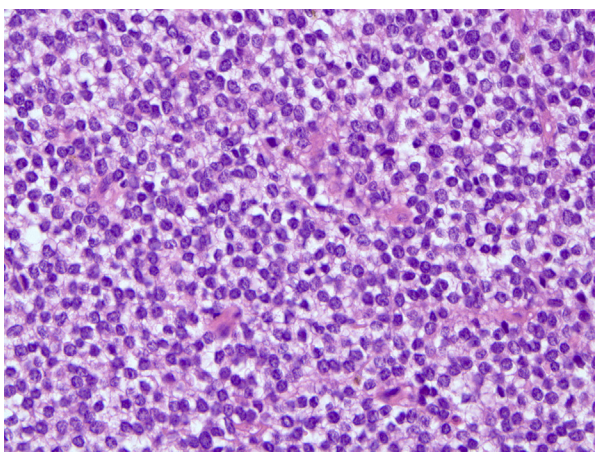

Figure 2. Histopathology slide demonstrating the typical appearance of ES/PNET. The lesion is composed of sheets of monotonous-appearing small, round blue cells with vacuolated/clear cytoplasm admixed with numerous small capillaries.

We conducted an extensive literature search and identified 19 vulvar and vaginal cases of ES (Table 1). The majority of these cases involve young women; all but two patients were younger than 40 years (median age 27, range 10 to 52 years). The average size of the tumor was 5.8 cm (SD ± 5.03 cm). Three cases presented with metastatic disease.

The differential diagnosis for a vulvar/vaginal mass most commonly includes benign lesions. A vaginal cyst may be an epidermal inclusion cyst. When a cyst is located in the proximal third of the vagina, it is often a Gartner duct cyst. A cyst in the periurethral region is likely a Skene's gland cyst or an abscess.[23] A cyst located in the distal, posterior vagina is most commonly an inflammatory process of a Bartholin gland. Additionally, a firm vaginal mass may be an extrauterine leiomyoma or lipoma. Furthermore, a cyst in the inguinal region could be a hydrocele in the canal of Nuck, which develops due to incomplete obliteration of peritoneal evagination anterior to the round ligament.[24] Sexually transmitted infections, such as genital herpes, donovanosis (granuloma inguinale), and chancroid, should also be considered as part of the diagnosis, particularly in the presence of an ulcerated lesion.

While the differential diagnosis for vulvar and vaginal lesions must always include primary vaginal or vulvar cancer, vulvar carcinoma rarely presents as a mass. Lymphoma and primary and metastatic breast cancer have been reported in the vulvar area.[25,26] There are no specific recommendations regarding imaging when evaluating vulvar masses, as most are presumed to be benign prior to resection. Imaging modalities, such as ultrasound, may be helpful to differentiate between a cystic and a solid structure and, therefore, aid in making the differential diagnosis. Such imaging results, however, are unlikely change the management or the surgical approach. Definitive diagnosis requires surgical excision and pathological evaluation. Imaging studies, such as CT and PET-CT, are instrumental in evaluating metastases and planning treatment.

Histological examination typically demonstrates a monomorphic population of small, round blue cells. Several neoplasms share these histological characteristics, including rhabdomyosarcoma, neuroblastoma, lymphoma, small cell carcinoma, melanoma, cutaneous adnexal tumors, and