Poster #25 | Primary vulvar Ewing sarcoma: a case report and systematic literature review

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Topic: Vulvar

Objectives
Ewing sarcoma is a round cell sarcoma genetically characterized by translocations of the FET gene family (typically the EWSR1 gene) to the ETS gene family (most commonly FLI1 or ERG). Infrequently, Ewing sarcoma may present as a primary vulvar tumor. Here we present a rare case of primary vulvar Ewing sarcoma in a post-menopausal patient. We also describe the clinical, pathological, and prognostic characteristics of the published cases of vulvar Ewing sarcoma identified in the medical literature.

Methods
A database search including MEDLINE (PubMed), Scopus, Cochrane (Wiley), and Google Scholar was performed using combinations of the keywords vulva, gynecologic, Ewing sarcoma, Ewing, primitive neuroectodermal tumor, and PNET. Articles not available in English were excluded. Systematic tabulation of clinically relevant data from each report was performed.

Results
A 74-year-old patient presented to the office with an asymptomatic, incidentally identified 5 cm vulvar mass. Her history was significant for FIGO stage I grade 2 endometrioid endometrial adenocarcinoma at age 50, status post staging hysterectomy, adjuvant external beam radiation and vaginal brachytherapy. Pathology from a biopsy of the vulvar mass demonstrated a round cell sarcoma. By immunohistochemistry, there was strong membranous expression of CD99 and by fluorescence in situ hybridization (FISH), EWSR1 gene rearrangement was demonstrated. Single gene next-generation sequencing confirmed an EWSR1-FLI1 gene fusion, consistent with Ewing sarcoma. Positron emission tomography identified no distant disease. She is currently undergoing treatment with vincristine, adriamycin, and cyclophosphamide, with planned surgical resection after 3 cycles of chemotherapy. Literature review identified 34 additional cases of primary vulvar Ewing sarcoma reported from Asia (15), North America (9), Europe (7), Africa (2), and Australia (1). Median age at diagnosis was 21.5 years with a range from 3 to 65 years, with our case representing the oldest reported age of presentation. Only 15% of reported cases occurred in post-menopausal patients and no other patients had a prior history of malignancy. Most tumors presented as a painless mass with median tumor size 5 cm (range 1 - 20 cm). Thirty-two of 34 cases reported positive CD99 immunohistochemistry, and EWSR1 gene rearrangement was confirmed by FISH in 30% and by reverse transcriptase-polymerase chain reaction (RT-PCR) in 15% of cases. No case of local invasion to surrounding structures was reported, although 30% of patients presented with metastasis at the time of diagnosis. Most common metastatic sites were pelvic lymph nodes (15%) and lungs (15%). Most patients were treated with excision (82%) and chemotherapy (76%) with 30% of patients also receiving radiation. Mean follow up was 17 months, during which 44% of patients experienced progressive or recurrent disease, with pulmonary (18%) and local (12%) recurrences reported most commonly. 53% of patients were alive at last follow-up.

Conclusions
Primary vulvar Ewing sarcoma is a rare vulvar sarcoma that typically presents in adolescents and patients
of reproductive age but may also present in post-menopausal patients. Vulvar Ewing sarcomas show typical CD99 expression and characteristic genetic translocations when tested. Ewing sarcomas are aggressive tumors with frequent pulmonary metastases, and nearly half of patients progress and die of disease at a mean follow-up time of less than two years. Ewing sarcoma should be included on the differential for vulvar sarcomas and treated promptly with multimodal therapy.

Abstract Table or Graph

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