Primary vulvar Ewing sarcoma / Peripheral primitive neuroectodermal tumor with pelvic lymph nodes metastasis: A case report

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Introduction:

Ewing sarcoma and peripheral primitive neuroectodermal tumors (PNET) are a group of soft tissue tumors that generally affect the bones. Although they are known to be aggressive tumors, but they are highly chemosensitive. Extraosseous Ewing sarcoma / PNET has been reported in few cases, less than 30 of them are in the vulva.

Case Study

A 27 years old female, presented to us with a 6x4 cm right subcutaneous solid vulvar lesion fixed to the underlying planes, causing pain and discomfort. The lesion has developed over the last 4 months.

Multiple biopsies showed small round cell tumor, abundant necrosis, and absence of architecturally distinct growth patterns.

Immunohistochemistry staining showed strong positivity for CD99 and vimentin, favoring the diagnosis of Ewing sarcoma / PNET.

MRI showed a 6 cm lesion in the right vulvar region extending deep and laterally, with close proximity to the muscles of the right lower extremity. Enlarged bilateral inguinal and right external iliac lymph nodes were also reported. PET/CT showed hypermetabolic enlarged inguinal lymph nodes bilaterally, and right pelvic lymph nodes, indicating a stage IVB disease.

After a multidisciplinary discussion, decision was taken to give neoadjuvant chemotherapy and decide on the extent of the surgery based on the response to chemotherapy and the residual disease volume afterwards.

The patient received 3 cycles of Vincristine, Adriamycin, Cyclophosphamide (VAC) alternating with Ifosfamide and Etoposide (IE). PET/CT was repeated after completion of treatment, and showed complete response to chemotherapy with no uptake in the inguinal or pelvic area. On clinical exam, there was a 2x1.5 cm residual tumor at the lower right vulva.

The patient underwent radical local excision of the right vulva with advancement flap. Residual tumor measured 1.6 cm and all margins were free of tumor, with the closest margin being the deep margin, 4 mm away from tumor. She then received 4 additional cycles of adjuvant chemotherapy (total of 7 cycles) and 30 sessions of external beam radiation therapy. She completed the full treatment 12 months after the initial diagnosis.

The patient is currently alive without disease 15 months after the initial diagnosis.

Discussion

Vulvar Ewing sarcoma / PNET has been rarely reported with less than 30 cases found in the literature, of which only 3 cases had presented with metastatic disease. Furthermore, no cases with metastasis to the pelvic lymph nodes were ever reported. Better prognosis was reported in pelvic cutaneous and superficial extraosseous ES / PNET as in our patient, in contrast with the deeper tumors. Due to the unclear treatment recommendations in vulvar ES, our treatment plan mirrored the one of skeletal ES / PNET.

Conclusion

ES / PNET are exceedingly rare in the female genitourinary tract; Nonetheless, they should be considered as part of the differential diagnosis in patients with vulvar lesions. Accurate pathologic diagnosis and a multidisciplinary approach are of utmost importance, in order to provide early and appropriate treatment.