Primary Ewing's Sarcoma of Penis – A Rare Case Report

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Introduction

Ewing's sarcoma (ES) and primitive neuroectodermal tumor (PNET) were considered as two ends of a spectrum of same tumor entity based on their similar clinical, immuno-histochemical and cytogenetic features. Extraskelatal Ewing's sarcoma (EES) is a type of Ewing's sarcoma that arises in soft tissue and is regarded as a member of the Ewing sarcoma/primitive neuroectodermal tumor (PNET) family. Primary visceral ES/PNETs are very rarely seen aggressive tumors. We present this case as this is forth case of ES/PNETs with penile localization [1–3]. One case of EES metastasizing to penis was also reported [4].

Case Report

A 29-year-old male patient presented in June 2008, with complaints of painful erection of penis every night and a mass at the base of penis for 2 years. His physical examination revealed approximately 8 cm large hard mass at the base of penis and two small separate nodules in the shaft of penis. There were no other relevant symptoms or signs. The

biopsy from mass was 'undifferentiated small round cell tumor'. Tumor cells expressed mic-2, but EMA and cyto-keratin were focally positive, and negative to desmin, myogenin & HMB 45 on immuno-histochemistry, giving the final diagnosis as PNET. The MRI scan of pelvis showed 9.1×7.1×6.2 cm, irregular, heterogenous mass, involving both corpora cavernosa, reaching near root of penis. Lesion was predominantly hyper-intense on T2W images and iso-intense on T1W images. Another hyper-intense lesion was noted in close relation with right corpora cavernosa at dorsal aspect of penis. The CT scan of thorax showed few rounded metastatic nodules in both lungs. Rest all work-up including bone scan and bone marrow biopsy were normal (Fig. 1).

We decided to use combined modality treatment with intent to cure. Anterior multi-agent chemotherapy was given including vincristine, adriamycin, cyclophosphamide, etoposide and ifosfamide. Local radiotherapy was given as 30 grays in 10 fractions. His bilateral pulmonary metastasectomy was done (in Mar 09) followed by total amputation of penis with perineal urethrostomy. In histo-pathology study viable tumor was seen and reported as Ewing's sarcoma. Lympho-vascular permeation was absent.

PET-CT scan done 2 months post-operatively showed metabolically active disease in the region of root of penis, lungs and mediastinal lymph nodes. Adjuvant chemotherapy was started using combination of vincristin, adriamycin, cyclophosphamide, topotecan and cisplatin. Follow-up CT scans showed increase in size of lung metastasis and local recurrence. It was considered as refractory Ewing's sarcoma. Imatinib was started followed by gefitinib but there was no response. Patient was complaining of severe local pain and respiratory distress. He was last seen in Dec. 2009. Despite of metastatic disease at presentation we could offer him a survival of about 18 months with the help of combined modality treatment.

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Fig. 1 a & b – Histo-pathologic view of specimen

Discussion

Ewing's sarcoma, or primitive neuroectodermal tumour (ES/PNET), is a neoplasm of undifferentiated small round cells that occurs in children, adolescents and young adults. EES/PNET have equal frequency in both males and females. The sites most commonly involved by EES/PNET are the extremities. Other common sites are the head and neck region, paravertebral region, and pelvis. It has also been documented in the pancreas, vagina, recto-vaginal septum, small bowel, prostate, ovaries, esophagus, and kidney [5].

MRI is considered the imaging modality of choice, being better than CT scan for demonstrating the pattern of tumor extension and for delineating the soft tissues. On MRI, the tumor is usually iso-intense to the muscles on T1W and hyper intense on T2W images, with enhancement on post contrast scans. The disease follows an aggressive course with a high recurrence rate. The presence of a distant metastasis is also common.

The EES/PNET is indistinguishable from the bony sarcomas on microscopy. EES is defined as having small homogeneous, round or oval cells containing solid material separated by fibrous septa. The cytoplasm of EES cells is rich in glycogen. The ultimate diagnosis should be based on both histology and immuno-histochemistry [5]. It has been reported that in patients with Ewing's sarcoma, immunohistochemical staining is positive for CD99 and CD56 but is negative for S-100, desmin, factor 8 cytokeratin and neurofilament as well as for T-cells, B-cells, LCA, CD 43 and CD68. Although both EES and PNET show expression of HBA-7 and the t11:22 translocation, only ESS contains PAS-stained glycogen in the cytoplasm. Expression of the *MIC2* gene is path-gnomonic for Ewing's sarcoma, but it may also occur in some cases of T-lymphoblastic lymphoma, poorly differentiated synovial sarcoma, small cell osteosarcoma and rhabdomyosarcoma. Ewing's sarcoma can be definitively diagnosed by detection of the *EWS* gene and rearrangement of ETS-related cancer genes [6].

The results of surgery alone for EES are poor in most cases, while patients receiving multimodal chemotherapy and radiotherapy have a much better prognosis [5]. Thus, an aggressive treatment protocol is needed. Excision of the tumor should be as radical as possible to minimize the tumor mass and to increase the effectiveness of adjuvant therapy. Multidrug chemotherapy is more effective than single-drug chemotherapy for the treatment of Ewing's sarcoma. It has been demonstrated that preoperative chemotherapy with the use of a combination of vincristine, doxorubicine, cyclophosphamide and etoposide allows for conservative surgical resection in selected cases with subsequent improvement of the postoperative condition [6].

Major prognostic factors include the absence or presence of a metastasis, tumor size, the extent of necrosis, the initial response to chemotherapy and the presence of EWS/FL 11 fusion transcripts [6].

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