Number and Title: 11159—Pelvic Ewing Sarcoma Survival in The Setting of Surgery With Radiation

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Background:

Pelvic Ewing sarcoma (ES) has poorer outcomes than extremity-based lesions. Methods of local control are controversial and there is a paucity of conclusive evidence for radiation alone, surgery alone, or a combination of both.

Purpose:

The purpose of this study was to evaluate the effect of different variables on survival and local control of patients with ES of the pelvis.

Methods:

A retrospective review was performed for all patients with a primary ES of the pelvic or sacral bones treated at a single institution between 1990-2013 (n=46). The primary endpoints were disease-specific mortality vs. survival with no evidence of disease (NED) at latest follow-up. Patients were excluded if follow-up was ≤1 year after completion of adjuvant chemotherapy (7, all NED), if there was an unplanned delay >6 weeks or omission of chemotherapy or local control (5, none NED), or if clear metastatic progression of disease during induction chemotherapy necessitated palliation treatment (2, both deceased). All patients received vincristine, adriamycin, cyclophosphamide, ifosfamide, and etoposide chemotherapy. Radiation doses were at least 4500 cGy for neo-/adjuvant and 5500 cGy for radiation-only regimens.

Results:

Of 32 patients with primary pelvic ES (mean age, 18.6 yrs; range, 5.8-57.0), 17 (53%) had NED at latest follow-up (122 mos; 37-308) and 15 (47%) succumbed (45 mos; 12-144). Twenty-one (65%) patients underwent surgery plus radiation compared to surgery alone (3/32, 10%) and radiation alone (8/32, 25%). Fifteen (47%) presented with localized disease and 17 (53%) with metastases (34% lung-only, 13% bone or bone marrow only, 6% multisystem).

Eighty-six percent (13/15) of patients who presented with localized lesions were survivors compared to only 24% (4/17) with metastases. None who presented with bone or bone marrow metastases survived (0/5) compared to 27% (3/11) with lung-only metastases. Of patients with histological analysis of the primary lesion, 68% (15/22) had ≥99% necrosis which was associated with 66% (10/15) survival. Two of the 4 patients with <90% necrosis survived. Twenty percent (2/10) of patients with primary lesions involving more than 1 contiguous pelvic area (31%, 10/32) such as sacroiliac, ilioacetabular or entire hemipelvis survived.

The use of surgery with radiation was associated with improved survival compared to either modality alone on univariate analysis (p=0.024), but 75% (6/8) of patients who received radiation alone had initial metastases compared to 42% (9/21) for surgery plus radiation. Involvement of only 1 pelvic area was also significant for improved survival (68%; p=0.021). Including other variables such as age at diagnosis, ≥99% necrosis, initial SUV (6.9; 2.1-13.7) or improvement of SUV (39%; 17%-100%) on PET scan, time to post-operative resumption of chemotherapy (0.9 mos, 0.7-1.6), and sites of metastasis, only the absence of metastatic disease (at presentation) was significant on both univariate and multivariate analysis (p=0.002).

The incidence of isolated local relapse was 0% (0/21) for surgery with radiation and 12% (1/8) for radiation alone. Seventeen percent (3/18) of patients survived after relapsed disease (1 isolated local recurrence,

1 bone-only metastasis, 1 lung-only metastasis). All three patients underwent metastatectomy and received high-dose chemotherapy and autologous stem-cell transplant. None of the patients who succumbed had an isolated pelvic recurrence. All relapsed with diffuse lung disease or multisystem involvement.

Conclusion:

For primary pelvic ES, approximately half of all patients have localized disease at presentation and the absence of metastatic involvement is the strongest predictor for survival. It is well-known that chemotherapy is key to survival, but a complete response is neither a guarantee nor an absolute necessity for survival. Up to one-third of patients die despite an excellent histologic response (≥99% necrosis) and approximately 30% of patients with lung metastases survive. The interplay between therapeutic modalities is complex, and treatment of the primary lesion has an integral role in patient survival.

The optimal approach for primary pelvic ES remains poorly defined. The majority of patients in this study underwent surgery with radiation and the minority that received radiation alone represented a higher-risk group. Treatment with surgery plus radiation allowed survival in 83% (10/12) with localized pelvic ES, 43% (4/7) with lung-only metastases, 25% (1/4) with primary lesions involving two contiguous pelvic areas, and 50% (2/4) with suboptimal (<90%) histologic response.

Level of Evidence: III, case-control or retrospective cohort study