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Ndamase, Ncumisa

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Local Control of Ewing Sarcoma/peripheral Primitive Neuroectodermal Tumour using multimodality treatment approach in children and adolescents in Johannesburg: A retrospective review

Dr. Ncumisa Ndamase
Radiation Sciences, Division of Radiation Oncology, School of Clinical Medicine University of the Witwatersrand, Johannesburg, South Africa

Student No: 1595963

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DECLARATION

I, Ncumisa Ndamase, declare that this thesis is my own work. It is being submitted for the degree of Master of Medicine in Radiation Oncology at the University of the Witwatersrand, Johannesburg. It has not been submitted for any degree or examination at this or any other university.



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Dr. Ncumisa Ndamase

20th day of February 2023 in Johannesburg

DEDICATION

This work is dedicated to my husband and children, who served as solid support system throughout my training and countless compromises that they have endured until the end. It is with gratitude I walked this journey with you all.

Publications and presentations
This work has not been published or presented at the conference.

ABSTRACT

Background

Ewing sarcoma and peripheral primitive neuroectodermal tumours (ES/pPNET) are aggressive bone and soft tissue malignancies that are most common in children and adolescents. The use of a multimodality treatment approach improves outcome especially following advancement in systemic chemotherapy. Local treatment strategies for local disease control continues to show promise in improving overall survival and local control rates in patients with localized disease.

Objective

The objective of this study was to determine the effectiveness of treatment in ES/pPNET by comparing surgery and radiotherapy in overall disease and event free survival.

Materials and Methods

This was a retrospective analysis of medical records of ES/pPNET patients treated in a public academic institution in Johannesburg. Records of fifty-four (54) patients with localized tumours treated from January 2000 to December 2015 were included in the study. Five-year overall survival and event-free survival as well as incidence of recurrence were estimated using the Kaplan-Meier for the analyzed cohorts. Cox regression models were used for analysis of prognostic factors and estimating odd ratios with 95% confidence intervals.

Results

The estimated 2-year and 5-year overall survival (OS) was 88.9% (95% CI: 0.7693 – 0.9485) and 77.7% (95% CI: 0.6400 – 0.8664) respectively. Based on the local treatment received, the 5-year OS was 73.3% for surgery only, 83.3% for radiotherapy only and 76.0% for patients that had both surgery and radiotherapy

In total, 18 (33.3%) of the 54 patients experienced recurrence; isolated distant recurrence occurred in 15 patients, the commonest site being the lungs (55.6%), followed by the

spine (14.8%) and the pelvis (14.8%). Combined local and distant recurrence occurred in 3 patients. The site of local recurrence in these three (3) was the pelvis. The estimated 5-year event free survival (EFS) for all patients was 65.8% (95% CI: 0.5130 – 0.7691). The estimated 5-year EFS was 63.6% (95% CI: 0.2042 – 0.8045) for surgery only, while for radiotherapy only, 66.7% (95% CI: 0.3771 – 0.8234) and for both surgery and radiotherapy 66.2% (95% CI: 0.4347 – 0.8149). No patient in this cohort of ES/pPNET had isolated local recurrence or failure following local treatment.

Patients with tumour size ≥ 8cm are 2.54 times likely to have local or distant tumour recurrence compared to patients with tumours < 8cm (OR: 2.54, 95% CI: 1.05-6.13, p=0.019). Patients with high pretreatment lactose dehydrogenase (LDH) level (at diagnosis), had approximately 5 times increased odds of recurrence compared with those with normal LDH (OR: 4.59, 95% CI: 1.34-15.74, p=0.010). The factors associated with recurrence were tumour size (OR: 2.64, 95% CI 1.08-6.45) and LDH (OR: 4.01, 95% CI 1.15-13.99). For mortality, LDH level (p<0.001) and disease recurrence (p<0.001).

Conclusion

Overall disease control in ES/pPNET is comparable for patients treated with surgery or definitive radiotherapy with chemotherapy. The risk of local failure is commoner in patients treated with definitive radiotherapy than surgery. Although radiation therapy is frequently applied in unfavorable disease group, local control outcomes are good and in many cases similar to surgical treatment outcomes. Distant failures account for the majority of relapses in this disease; therefore there is need for better and improved systemic therapies for both local and distant disease control.

Keywords: ES/PNET, surgery, radiotherapy (RT), disease recurrence, overall survival (OS), event free survival (EFS), lactact