Extraskeletal Ewing Sarcoma- Experience in a Tertiary Cancer Care Centre of India

Authors: Himanshu Rohela

Abstract: BACKGROUND: Ewing sarcoma can arise in either bone or soft tissue. Extraskeletal Ewing sarcoma (EES) is an uncommon primary tumor of the soft tissues, accounting for 20 30% of all reported cases of ES. AIM: Was to investigate demographic distribution, survival analysis and factors affecting the survival and recurrence in patients of EES. METHODS: Retrospective study of 19 biopsy-proven EES was performed. Overall survival (OS) using log-rank test and factors affecting OS and local recurrence (LR) were evaluated for the entire cohort. RESULTS: Patients with EES had a mean age of 19.5 and it was more commonly seen in males (63%). Axial location (58%) and solitary presentation (84%) were more common. The average size was 11 cm, 3 of 19 were metastatic at presentation, with the lung beings the most common site for metastasis. 17 received NACT, 16 with VAC-IE regimen and 1 underwent a second line with GEM/DOCE regimen. Unplanned surgery was done in 2 of 19. 3 patients received definitive RT and 13 underwent surgical-wide local excision. 2 of 13 showed good response to NACT. 10 patients required readmission out of which 6 patients had chemotherapy-related complications, 2 had surgical site complications and one patient developed secondary AML post-completion of treatment. A total of 4 patients had a recurrence. One had local recurrence alone, one had distant recurrence alone and 2 patients had a distant and local recurrence both. Tumor size >10 cm, axial location, and previous unplanned surgery was associated with higher LR rate. The mean overall survival was 32 months (2.66 years), with higher rates seen in non-metastatic and non-recurrent settings. CONCLUSIONS: Early and accurate diagnosis is the key to the management of EES, with promising results seen via NACT and RO resection regimens. But further studies with larger study groups are needed to standardize the treatment protocol and evaluate its

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