Title
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The role of chemotherapy and radiotherapy in localized extraskeletal osteosarcoma

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Abstract

Purpose: The role of chemotherapy (CT) and radiotherapy (RT) for management of extraskeletal osteosarcoma (ESOS) remains controversial. We examined disease outcomes for ESOS patients and investigated the association between CT/RT with recurrence and survival.

Patients and methods: Retrospective review at 25 international sarcoma centers identified patients ≥18 years old treated for ESOS from 1971 to 2016. Patient/tumour characteristics, treatment, local/systemic recurrence, and survival data were collected. Kaplan-Meier survival and Cox proportional-hazards regression and cumulative incidence competing risks analysis were performed.

Results: 370 patients with localized ESOS treated definitively with surgery presented with mainly deep tumours (n = 294, 80%). 122 patients underwent surgical resection alone, 96 (26%) also received CT, 70 (19%) RT and 82 (22%) both adjuvants. Five-year survival for patients with localized ESOS was 56% (95% CI 51%-62%). Almost half of patients (n = 173, 47%) developed recurrence: local 9% (35/370), distant 28% (102/370) or both 10% (36/370). Considering death as a competing event, there was no significant difference in cumulative incidence of local or systemic recurrence between patients who received CT, RT, both or neither (local p = 0.50, systemic p = 0.69). Multiple regression Cox analysis showed a significant association between RT and decreased local recurrence (HR 0.46 [95% CI 0.26-0.80], p = 0.01).

Conclusion: Although the use of RT significantly decreased local recurrences, CT did not decrease the risk of systemic recurrence, and neither CT, nor RT nor both were associated with improved survival in patients with localized ESOS. Our results do not support the use of CT; however, adjuvant RT demonstrates benefit in patients with locally resectable ESOS.

Keywords: Chemotherapy; Extraskeletal osteosarcoma; Radiation therapy; Radiotherapy; Soft-tissue osteosarcoma.