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Management of Ewing sarcoma family of tumors: current scenario and unmet need

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Abstract

Ewing sarcoma family tumors (ESFT) are heterogeneous, aggressive group of disease with peak incidence in adolescent and young adults. The outcome has been improved dramatically from 10% with surgery and radiotherapy alone to 65-70% now, in localized disease, with the introduction of chemotherapy. Chemotherapy regimen evolved from single agent to multiagent with effort of many cooperative clinical trials over decades. The usual treatment protocol include introduction of multi-agent chemotherapy in neoadjuvant setting to eradicate systemic disease with timely incorporation of surgery and/or radiotherapy as local treatment modality and further adjuvant chemotherapy to prevent recurrence. Risk adapted chemotherapy in neoadjuvant and adjuvant setting along with radiotherapy has been used in many international collaborative trials and has resulted in improved outcome, more so in patients with localized disease. The role of high dose chemotherapy with stem cell rescue is still debatable. The outcome of patients with metastatic disease is dismal with long term outcome ranges from 20%-

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