Malignant ectomesenchymoma in children and adolescents: report from the Cooperative Weichteilsarkom Studiengruppe (CWS).

Abstract:
Malignant ectomesenchymoma (MEM) is a soft tissue tumor with heterologous rhabdomyoblastic components believed to arise from pluripotent migratory neural crest cells. To date merely 50 cases have been published and the knowledge about the course of disease and optimal treatment is limited. Six patients with MEM were registered 1996-2009. The diagnosis was confirmed according to current criteria. Their treatment and outcome was analyzed. The median age of the three females and three males was 0.6 years (range, 0.2-13.5). The mesenchymal component in all tumors was rhabdomyosarcoma (RMS), the neural component ganglioneuroblastoma/neuroblastoma (n = 5) and peripheral primitive neuroectodermal tumor in one case. Five patients presented with localized, one with metastatic disease. All but one patient received multiagent chemotherapy during their initial treatment. The tumors of 4/5 patients with localized MEM were at least grossly resected at best surgery; the patient without gross resection was additionally irradiated. Three of four evaluable tumors responded well to induction chemotherapy. All patients achieved a first complete remission.
(CR), but three recurrences (two local, one systemic) occurred. The individual with metastatic MEM did not survive, but all five patients with localized MEM are currently alive in CR with a median follow-up of 5 years (range: 2.1-13.7). Risk-factors and outcome of MEM appear to be comparable with other highly malignant pediatric soft tissue sarcoma when a multimodal treatment strategy including chemotherapy and adequate local treatment is pursued. We propose that treatment of patients with MEM be done according to pediatric protocols similar to other rhabdomyosarcoma-like soft tissue sarcoma.

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