

MALIGNANT SOLID TUMORS IN CHILDREN WITH IMMUNODEFICIENCY SYNDROMES

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Objective: To evaluate our patients with immunodeficiency syndromes associated with malignant solid tumors, and to examine survival rates and prognosis with respect to type of immunodeficiency disease. **Patients and Methods:** Twenty two patients who were diagnosed with malignant solid tumors and immunodeficiency syndromes between years 1972 and 2003 were retrospectively analyzed. Age range was 1.5-24 years (median: 8), male/female ratio was 8/14. We had 12 (54.5%) patients with non-Hodgkin lymphoma (NHL), 8 (36.5%) with Hodgkin disease (HD), one (4.5%) with mucinous adenocarcinoma of the colon, and one (4.5%) with brain stem glioma. Fifteen (68.2%) patients had Ataxia-telangiectasia (AT), three (13.6%) had common variable immunodeficiency syndrome (CVID), 2 (9.2%) had Bloom syndrome, one (4.5%) had combined immunodeficiency, and one (4.5%) had selective immunoglobulin A deficiency. **Results:** Out of 15 patients with AT 9 patients had NHL, five had HD, and one had brain stem glioma. Two patients with CVID had NHL and one had HD. One of the patients with Bloom syndrome had HD and the other had colon carcinoma. The overall survival (OAS) in whole group was 24.1. One AT patient died after 10 years of remission without malignant disease. OAS in NHL, HD were 16.6%, 43.7%, respectively. OAS in AT patients was 20%. Two patients with CVID and NHL had died with disease progression. **Conclusion:** In our series, most of the patients had Ataxia-telangiectasia (68.2%). The survival rates of the malignant diseases were very poor in immunodeficiency. OAS in NHL patients was relatively worse than HD patients.

