ATAXIA-TELANGIECTASIA-ASSOCIATED PEDIATRIC HODGKIN LYMPHOMA: HIGH INITIAL CHEMOSENSITIVITY BUT DISMAL SURVIVAL

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INTRODUCTION

Ataxia-telangiectasia (AT) is a rare inherited disorder associated with immunodeficiency, defective DNA damage response, chromosomal instability, and marked radiosensitivity, predisposing affected children to lymphoid malignancies while substantially increasing treatment-related toxicity. Evidence guiding optimal management of Hodgkin lymphoma (HL) in children with AT remains limited, particularly regarding safe chemotherapy intensity adaptation and supportive care optimization.

METHOD

We retrospectively reviewed children with AT diagnosed with HL at Children's Cancer Hospital Egypt between July 2007 and July 2024. Data collected included clinical presentation, stage, B symptoms, risk group, extranodal disease, histopathology, frontline treatment, dose modifications, number of cycles delivered, interim response assessment, and survival outcomes.

CONCLUSIONS

AT-associated pediatric HL appears highly chemosensitive, but outcomes are dominated by extreme non-relapse mortality, mainly related to infectious, pulmonary, and treatment-related complications. These findings underscore the urgent need for AT-adapted treatment strategies that preserve efficacy while minimizing toxicity, together with intensive prophylaxis, pulmonary support, and immune optimization..

RESULTS

Fourteen patients were identified, with a median age at diagnosis of 9 years (range, 6-17 years); 11/14 were male. Advanced presentation was frequent, with stage IV disease in 7/14 patients (50.0%) and B symptoms in 6/14 (42.9%). Risk stratification showed 7/14 high-risk (50.0%), 5/14 intermediate-risk (35.7%), and 2/14 low-risk disease (14.3%). Histology was predominantly classical HL, with mixed cellularity in 8/14 (57.1%) and nodular sclerosis in 5/14 (35.7%); 1 case showed features intermediate between diffuse large B-cell lymphoma and classical HL. Frontline treatment was heterogeneous and toxicity-adapted: 4/14 patients (28.6%) received AVPC with 50% dose reduction, 3/14 (21.4%) AVPC with 25% dose reduction, 1/14 (7.1%) full-dose AVPC, and 4/14 (28.6%) ABVD. One additional patient (7.1%) received 2 cycles of ABVD with 50% dose reduction and omission of doxorubicin because of marked sensitivity, then shifted to COPDAC with 25% dose reduction. The patient with lymphoma showing intermediate features between diffuse large B-cell lymphoma and classical HL received rituximab alone because of very poor general condition. Eleven patients achieved complete metabolic response after 2 cycles, 1 had progressive disease, and 2 died before response evaluation. Despite the high early response rate, all patients ultimately died, predominantly from non-relapse causes, mainly chest infection, respiratory failure, and septic shock. Median survival was 6 months.

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REFERENCES

Sandoval C, Swift M. Hodgkin disease in ataxia-telangiectasia patients with poor outcomes. Med Pediatr Oncol. 2003;40(3):162–166. doi:10.1002/mpo.10251.

Niehues T, Schellong G, Dörffel W, Bucsky P, Mann G, Körholz D, et al. Immunodeficiency and Hodgkin's disease: treatment and outcome in the DAL-HD78–90 and GPOH-HD95 studies. Klin Padiatr. 2003;215(6):315–320. doi:10.1055/s-2003-45498