

Clinical, pathological and genetic features of primary mediastinal large B-cell lymphomas and mediastinal gray zone lymphomas in children.

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Source

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Abstract

Background Primary mediastinal large B-cell lymphoma is a rare lymphoma accounting for no more than 3% of all B-cell lymphomas in children and adolescents. However, patients in this young age group with this lymphoma have the shortest event-free survival of patients with any B-cell lymphoma under current standard chemotherapy protocols. Lymphomas with features intermediate between primary mediastinal large B-cell lymphoma and classical Hodgkin's lymphoma (mediastinal gray zone lymphomas) have been acknowledged in the latest World Health Organization classification. Recent studies suggest that mediastinal gray zone lymphomas have an aggressive clinical course whereas patients, at least adult ones, with primary mediastinal large B-cell lymphoma might respond very well to chemotherapy in combination with anti-CD20 antibody. **DESIGN AND METHODS:** We aimed to evaluate whether biological differences or so far unrecognized admixed mediastinal gray zone lymphomas might explain the relatively poor outcome of pediatric patients with apparent primary mediastinal large B-cell lymphoma. We, therefore, performed a retrospective histopathological, immunohistochemical and interphase cytogenetic analysis of 52 pediatric lymphomas. **RESULTS:** The childhood primary mediastinal large B-cell lymphomas (n=44) showed a similar pattern of histology, immunophenotype and gains at 9p (59%) and 2p (41%) as adult cases, as determined from published data. We identified only four so far unrecognized cases of mediastinal gray zone lymphoma among 52 lymphomas registered in previous trials. **Conclusions** Mediastinal gray zone lymphoma is very rare in children and adolescents. It does, therefore, seem unlikely that these lymphomas account for the unsatisfactory clinical results with current therapy protocols in pediatric patients. These data have major implications for the design of future treatment protocols for mediastinal lymphomas in children and adolescents.

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