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Epstein-Barr virus-positive systemic NK/T-cell lymphomas in children: report of six cases.

Rodríguez-Pinilla SM, Barrionuevo C, García J, de los Ángeles M, Pajares R, Casavilca S, Montes J, Martínez A, Montes-Moreno S, Sánchez L, Piris MÁ.

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Abstract

The World Health Organization lymphoma classification recognizes two different Epstein-Barr virus (EBV)-positive T-cell lymphoproliferative disorders of childhood: systemic EBV-positive T-cell lymphoproliferative disease of childhood, and hydroa vacciniforme-like lymphoma, which is more prevalent in Asia and Latin America. The aim of this study was to characterize six cases of paediatric EBV-positive peripheral T-cell lymphoma with distinct features. All cases were male, with a median patient age of 9 years (range: 5-17 years). Most of them presented suddenly with fever, weight loss, hepatosplenomegaly, peripheral lymphadenopathy, and high lactate dehydrogenase (LDH) levels. Moreover, gut, lung or soft tissues of the abdominal wall were also affected in four cases. Partial to total replacement of the lymph node by pleomorphic infiltration of atypical neoplastic cells was found in all cases. Vasculitis and geographical areas of necrosis were seen in three and four cases, respectively. Neoplastic cells showed expression of EBV-encoded RNA, T-cell markers (CD2 and CD3), and cytotoxic markers (TIA1, granzyme-B, and perforin). CD56 and T-cell receptor -γ were expressed in one case each. TCR-BF1, CD4, CD8 and anaplastic lymphoma kinase were negative. In all cases, the disease progressed rapidly, causing death of the patient, with a median survival of 7.1 months (range: 1-13 months). These cases probably represent a solid form of systemic EBV-positive T-cell lymphoproliferative disease of childhood, which requires identification and the development of appropriate therapy.

First-line treatment for primary testicular diffuse large B-cell lymphoma with Rituximab-CHOP, CNS prophylaxis, and contralateral testis irradiation: final results of an international phase II trial.

Vitolo U, Chiappella A, Ferreri AJ, Martelli M, Baldi I, Balzarotti M, Bottelli C, Conconi A, Gomez H, Lopez-Guillermo A, Martinelli G, Merli F, Novero D, Orsucci L, Pavone V, Ricardi U, Storti S, Gospodarowicz MK, Cavalli F, Sarris AH, Zucca E.

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Abstract

PURPOSE: Primary testicular lymphoma (PTL) has poor prognosis with failures in contralateral testis, CNS, and extranodal sites. To prevent these events, we designed an international phase II trial (International Extranodal Lymphoma Study Group 10 [IELSG- 10]) that addressed feasibility and activity of conventional chemoimmunotherapy associated with CNS prophylaxis and contralateral testis irradiation. The trial was conducted by the IELSG and the Italian Lymphoma Foundation. PATIENTS AND METHODS: Fifty-three patients (age 22 to 79 years) with untreated stage I or II PTL were treated with six to eight courses of rituximab added to cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) every 21 days (RCHOP21); four doses of intrathecal methotrexate (IT-MTX) and radiotherapy (RT) to the contralateral testis (30 Gy) for all patients and to regional lymph nodes (30 to 36 Gy) for stage II disease. RESULTS: All patients received R-CHOP21, 50 received CNS prophylaxis, and 47 received testicular RT. With a median follow-up of 65 months, 5-year progression-free survival and overall survival rates were 74% (95% CI, 59% to 84%) and 85% (95% CI, 71% to 92%), respectively. Ten patients relapsed or progressed: two in lymph nodes, five in extranodal organs, and three in the CNS. The 5-year cumulative incidence of CNS relapse was 6% (95% CI, 0% to 12%). No contralateral testis relapses occurred. Ten patients died: lymphoma (n = 6), secondary leukemia (n = 2), heart failure (n = 1), and gastric cancer (n = 1). Grade 3 to 4 toxicities were neutropenia, 28%; infections, 4%; and neurologic, 13%. No deaths occurred as a result of toxicity. CONCLUSION: This international prospective trial shows that combined treatment with R-CHOP21, IT-MTX, and testicular RT was associated with a good outcome in patients with PTL. RT avoided contralateral testis relapses, but CNS prophylaxis deserves further investigation.