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The primary cutaneous CD30- positive lymphoproliferative diseases; the own observations of the center, prospects of therapy

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Primary cutaneous T-cell lymphomas are the heterogeneous group of T-cell lymphoproliferative diseases (LPD), which are developed mainly in the skin and characterized by diagnostic features, clinical course and therapeutic approach. Primary cutaneous anaplastic lymphoma (c-ALCL) and lymphomatoid papulosis (LyP) are diagnosed In a quarter of cases of all T-cell skin lymphomas. The tumor cells in primary cutaneous CD30-positive skin lymphomas express CD30 in more than 75% of cases. The majority of patients with cutaneous CD30+ LPD have an indolent course with a favorable prognosis, the resistant course of disease develops in about 30% of cases and fatal cases from lymphoma are registered in 8% of cases. Most commonly, the treatment of these forms of LPD includes the surgical removal, radiation therapy or small doses of methotrexate, the systemic chemotherapy is used to generalize the process. Recently, monoclonal antibodies have been included in clinical practice for the treatment of skin lymphomas, one of which is brentuximab vedotin, the use of which has shown a rather high efficiency in the treatment of CD30 + skin lymphomas: more than 2/3 of the patients responded to the treatment, despite the many lines of therapy (median of prior effect is 3.1 for c-ALCL).

Results

The general group of patients with cutaneous T-cell lymphomas and who received the consultative-diagnostic or inpatient treatment at the National Research Centre of Hematology includes 328 patients. Among them the CD30-positive cutaneous LPD were verified in 33 patients (10%): 16 patients with LyP, 17 patients with c-ALCL. The eruptions were regressed on its own without specific therapy in 75% of patients with LyP and 4 patients received the treatment. As well as one patient with the relapsing course of disease, with a long period of self-regression of the papules, with the lack of the treatment effect (phototherapy and low-dose methotrexate) received brentuximab vedotin (BV). The complete clinical response was achieved after the first cycle of monotherapy with BV and persists for more than a year.

Patients with c-ALCL more frequently needed the specific therapy (76% of patients), only 4 patients were observed with the self-regression of the papules within 6-9 weeks. Most patients received the interferon alfa therapy with the complete clinical response; the rest patients - local radiation therapy, the small doses of methotrexate. The systemic chemotherapy was done to the 5 patients. Considering the common process, however 2 of them died from infectious complications during the chemotherapy, 2 patients had early recurrence and only 1 patient is in the long-lasting complete remission after the high-dose of chemotherapy.

In summary, the primary cutaneous CD30 + lymphoproliferative diseases cover a spectrum of benign condition with the so-called “malignant” phenotype. LyP and c-ALCL have a favorable prognosis, 5-year survival rate is exceeding 95%, photo- and immunotherapy are successfully used in the treatment, while the use of systemic chemotherapy should be avoided (shortening of the remissions duration, progression of the infectious complications). The use of targeted therapy (brentuximab vedotin) increases the therapeutic ability in the situations where the routine treatment options are ineffective.

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