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OPTIMIZATION OF DIAGNOSTICS OF ANAPLASTIC LARGE CELL LYMPHOMA IN CHILDREN

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✓ Resume

The article analyzes the data obtained retrospectively from the case histories of 12 patients who were treated in the THF clinic of the RSNPMCORiR in the chemotherapy department with a diagnosis of ALCL, the average age was 9.8 years (3-14 years), the ratio between boys was 7 (58.3 %) and girls - 5 (41.6%) was 1.4:1. All patients underwent clinical (medical history, general examination), laboratory (general and biochemical blood tests, morphological and cytochemical studies of BM, morpho-immunological) studies of tumor tissue. In order to determine the prevalence of the tumor process, instrumental imaging methods were performed: ultrasound, CT, MRI, X-ray, scintigraphy. The clinical characteristics of the disease to determine prognostic risk groups in ALCL are affected by damage to the lung tissue, skin and bones. The results of the study indicate that given the small number of these lesions among the patients included in our study, 66.6% had a 2 prognostic risk group.

Key words: anaplastic large cell lymphoma, children, lung tissue, skin, bones.

BOLALARDA ANAPLASTIK YIG'IY HUYURALI LIMFOMALARNI DIAGNOSTIKASINI OPMOMALLASHTIRISH

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✓ Rezyume

Maqolada ALCL tashxisi bilan kimyoterapiya bo'limida RSNPMCOR THF klinikasida davolangan 12 bemorning kasallik tarixidan retrospektiv ravishda olingan ma'lumotlar tahlil qilinadi, o'rtacha yoshi 9,8 yosh (3-14 yosh), ularning nisbati. o'g'il bolalar 7 (58,3%) va qizlar - 5 (41,6%) 1,4: 1 edi. Barcha bemorlar klinik (tibbiy tarix, umumiy tekshiruv), laboratoriya (umumiy va biokimyoviy qon testlari, CM ning morfologik va sitokimyoviy tadqiqotlari, o'sma to'qimalarining morfo-immunologik) tadqiqotlaridan o'tkazildi. O'sma jarayonining tarqalishini aniqlash uchun instrumental ko'rish usullari amalga oshirildi: ultratovush, KT, MRI, rentgen, sintigrafiya. ACCL uchun prognostik xavf guruhlarini aniqlash uchun kasallikning klinik xususiyatlari o'pka to'qimalariga, teriga va suyaklarga zarar etkazishdir. Tadqiqot natijalari shuni ko'rsatadiki, bizning tadqiqotimizga kiritilgan bemorlar orasida ushbu lezyonlarning kam sonini hisobga olgan holda, 66,6% 2 prognostik xavf guruhiga ega edi.

Kalit so'zlar: anaplastik yirik hujayrali limfoma, bolalar, o'pka to'qimasi, teri, suyaklar.

ОПТИМИЗАЦИЯ ДИАГНОСТИКИ АНАПЛАСТИЧЕСКОЙ КРУПНОКЛЕТОЧНОЙ ЛИМФОМЫ У ДЕТЕЙ

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✓ Резюме

В статье проведен анализ данных полученных ретроспективно из историй болезни 12 больных, проходивших лечение в клинике ТГФ РСНПМЦОиР в отделении химиотерапии с диагнозом АККЛ, средний возраст составил - 9,8 лет (3-14 лет), соотношение между мальчиками - 7 (58,3%) и девочками - 5 (41,6%) составило 1,4:1. Всем больным проведены клинические (анамнез заболевания, общий осмотр), лабораторные (общий и биохимический анализы крови, морфологические и цитохимические исследования КМ, морфо-иммунологические) исследования опухолевой ткани. С целью определения распространенности опухолевого процесса проведены инструментальные методы визуализации: УЗИ, КТ, МРТ, рентген, сцинтиграфия. Клиническим характеристикам заболевания для определения прогностических групп риска при АККЛ имеет значение поражение легочной ткани, кожи и костей. Результаты исследования свидетельствуют, что учитывая немногочисленность этих поражений среди больных, включенных в наше исследование, 66,6% имели 2 прогностическую группу риска.

Ключевые слова: анапластическая крупноклеточная лимфома, дети, легочная ткань, кожа, кости.

Relevance

According to WHO (2012), over the past 10 years there has been an increase in the incidence of non-Hodgkin's lymphomas (NHL), which is up to 10% in the structure of all malignant neoplasms. A similar incidence rate is noted in the contingent of pediatric patients, which, according to the literature, is 5-8% of all malignant tumors [1,2]. In recent years, the incidence of NHL in children has increased [3,4]. Moreover, boys get sick approximately 3 times more often than girls. The disease is most common in patients aged 5-14 years. It should be noted that today the incidence of NHL among children in Uzbekistan tends to increase, so for the period 2011-2015. gross incidence rate in 2011. amounted to 0.40/000, and in 2015. - 0.70/000. [5] At the same time, until now there is no information on the prevalence of the disease by territorial, age and gender, which is possibly due to the lack of children's oncological departments in many regional oncological institutions and the difficulty of differential diagnosis of the disease with other hemoblastoses.

The development of the most common embryonic tumors is associated with structural changes in the chromosomal apparatus, in particular, with the deletion (loss) of certain parts of the chromosome, as a result of which the action of tissue-specific suppressor mechanisms is activated and, possibly, certain oncogenes are activated.

The mapping of suppressor genes in retinoblastoma, nephroblastoma, and neuroblastoma should be considered the most fundamental discovery in this area. These mutational changes can occur both in germinal (sex) cells (then they are considered hereditary and are transmitted in an autosomal dominant manner to offspring) and in somatic cells of the child (in these cases, the tumor is not inherited) [4]. For the transformation of a mutant cell into a cancer cell, the presence of another event, most often a mutation, in the same cell is necessary. The probability of the second event determines the penetrance (probability of manifestation) of the tumor. The first reports and descriptions of anaplastic large cell lymphoma (ALCL) were published more than 30 years ago [3]. With the development of oncomorphology and immunogenetics, this pathology has been studied in detail and at present there are descriptions of its cytogenetic and immunological features. The clinical map of ALCL is diverse, is characterized by the presence of nodal and extranodal lesions of organs and systems, morphological characteristics, and can be represented by classical, small cell and lymphohistiocytic variants. IHC analysis can determine the presence of T-cell markers, but even in their absence, the T-cell origin of ALCL is confirmed by genetic analysis with the determination of the T-cell receptor and cytotoxic T-cell proteins in tumor cells.

Purpose of the study. The aim of our work was to optimize the diagnosis of anaplastic large cell lymphoma in children.

Materials and methods

In this study, we analyzed the data obtained retrospectively from the case histories of 12 patients who were treated at the THF clinic of the RSSPMCORiR in the chemotherapy department with a diagnosis of ALCL, the average age was 9.8 years (3-14 years), the ratio between boys was 7 (58, 3%) and girls - 5 (41.6%) was 1.4:1

All patients underwent clinical (medical history, general examination), laboratory (general and biochemical blood tests, morphological and cytochemical studies of BM, morpho-immunological) studies of tumor tissue.

In order to determine the prevalence of the tumor process, instrumental imaging methods were performed: ultrasound, CT, MRI, X-ray, scintigraphy.

Results and discussion

An analysis of clinical manifestations showed that in patients in the study group, symptoms of intoxication (weakness, lethargy, sweating, weight loss > 10%, fever) were observed in 9 (75.0%) patients. From the anamnesis, it was found out that the first sign that parents paid attention to was an increase in 1 / y in 7 (58.3%) children, skin manifestations in the form of spots, plaques were determined in 4 (33.3%) children, a pain symptom in the bones - met infrequently in 2 (16.6%) patients. An accidental finding during ultrasound and X-ray studies revealed tumor formations in 3 (25.0%) patients.

The results of the study showed that the clinical picture of ALCL is characterized by the predominant involvement of peripheral lymph nodes and soft tissues - 58.3% and 50.0%, respectively. Rare cases of lesions were the spleen and lungs, then the skin - 33.3%, bones and abdominal organs - 25.0%, mediastinum - 16.6%. Isolated cases of damage were characteristic of the pancreas, stomach,

liver, testis, CNS . No CM lesions were found in any of the patients. In CNS lesions, the clinic was characterized by changes in neurological symptoms for the brain.



Figure 1. Frequency of l/n and organ damage in ALCL

Most often, in the group of patients studied, there was a lesion of two or more anatomical regions in 9 (75.1%) patients, which were mostly extranodal and occurred with the same frequency in both boys and girls.

On the basis of ultrasound and CT, the volume of the tumor lesion and size were determined, most often the tumors were from 5 to 10 cm in size - 56.3%, more than 10 cm - 16.6, up to 5 cm - 25.0%. Data on the distribution of patients depending on the size of tumor conglomerates suggest that ALCL is not characterized by the formation of large tumor conglomerates, since in most patients (56.3%) the size of the tumor was 5-10 cm. Primary extranodal lesions in ALCL occurred in 33.3%, with no significant differences by gender.

The lesions of the bones of the skeleton were presented in the form of specific infiltrates of the bone tissue, lower leg, shoulder, thigh and ribs. One patient had a lesion of the skull bones, which was characterized by a stormy, aggressive and rapid process with brain damage, i.e. with damage to the bones of the skeleton, there is a rapid spread of the process to the surrounding tissues and internal organs.

The clinical stage of the disease was determined on the basis of chest X-ray data, CT, ultrasound of the abdominal cavity and extraperitoneal zones in the presence of a lesion. If the process proceeded with damage to the bones of the skeleton, if necessary, scanning of bone structures was performed. The results of clinical data showed that the largest number of patients had generalized stage III-IV ALCL, while stage III. - 41.6%; IV-st. – 33.3%; patients in I-st. was not determined, with II-Art. – 25.0%. Given the aggressiveness of ALCL, the clinical manifestations of the disease have a fulminant form with a rapid spread and generalization of the process, which explains a large percentage of patients in stages III-IV.

When studying the level of LDH in the blood serum in order to determine the biological activity of lymphoma, LDH activity of more than 1000 U/l was observed only in 1 (8.3%) patient. Patients with LDH levels of 500–1000 U/l had the highest percentage, which indicates that this marker does not change in patients with ALCL, and the level of 500–1000 U/l corresponds to the norm in this pathology.

Conclusions

According to the clinical characteristics of the disease, damage to the lung tissue, skin, and bones is important for determining prognostic risk groups in ALCL. Considering the paucity of these lesions among the patients included in our study, 66.6% had a prognostic risk group 2.

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