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A RARE CASE OF INFLAMMATORY MYOFIBROBLASTIC ABDOMINAL TUMOR IN A CHILD

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The article presents a clinical case of a rare inflammatory myofibroblastic tumor of the abdominal cavity in a 6-year-old boy. The child had a high fever, laboratory tests revealed an increase in acute phase proteins, and according to instrumental studies - a large tumor of the abdominal cavity, without a clear organ affiliation. During laparoscopic revision, the possibility of total tumor removal was established; laparotomy was performed and the tumor was radically removed.

Keywords: rare tumors, children, inflammatory myofibroblastic tumor, ALK gene translocation.

Introduction. Inflammatory myofibroblastic tumor (IMT) is a rare tumor with uncertain biological behavior, characterized by heterogeneity both in its histological pattern and in the molecular genetic changes underlying its development [1]. The etiology and pathogenesis of IMT are not fully understood. Several risk factors have been described, including smoking, trauma, and systemic IgG4-associated sclerosing disease, and there are hypotheses suggesting an inadequate immunological response to tissue damage. Human herpes virus type 8 and Epstein-Barr virus are most often considered as etiological agents [6]. The diagnosis of IMT has long been a diagnosis of exclusion, with a broad differential diagnosis ranging from local inflammatory process and idiopathic retroperitoneal fibrosis to inflammatory fibrosarcoma. The identification of ALK gene rearrangements has made a significant contribution to the understanding of the mechanisms of treatment of this rare mesenchymal tumor.

Most often, this disease is described in childhood and adolescence, and is mainly localized in the lungs, abdominal cavity, retroperitoneal space and soft tissues of the pelvis. IMT has the character of local invasive growth, can recur, but rarely metastasizes [2, 3]. The clinical picture depends on the location of the tumor and is often accompanied by general symptoms of inflammation such as fever or malaise. Laboratory manifestations of IMT include leukocytosis, increased levels of acute phase proteins: C-reactive protein, ESR, fibrinogen. The X-ray appearance of IMT is nonspecific and is often interpreted as malignant neoplasms. Histological examination is decisive in making the correct diagnosis [5].

At the molecular level, approximately half of IMTs contain a clonal cytogenetic aberration, a translocation of the ALK gene, which makes it possible to differentiate IMTs from other spindle cell tumors in children and is a target for inoperable and recurrent cases. Radical surgery is the basis in the treatment of IMT [1].

Overall, retrospective studies have shown that IMT has a favorable prognosis. The 5-year event-free survival (EFS) and overall survival (OS) rates are 82.9% and 98.1%, respectively [7].

Purpose of the study: To describe a rare case of inflammatory myofibroblastic tumor in the abdominal cavity in a child.

Materials and methods. A retrospective analysis of the medical records of a patient who was in the oncohematological and surgical departments of the Pediatric center of the State Autonomous Institution of the Republic of Sakha (Yakutia) "Republican Hospital No. 1- NCM named after M.E. Nikolaev". A full in-depth examination was carried out in the oncohematology department. Laboratory tests (general blood and urine analysis, biochemical blood test, study of the coagulation system) and instrumental studies (ultrasound of the abdominal organs, ECG, computed tomography of the chest and abdominal organs, MRI of the abdominal organs with contrast enhancement) were performed. A histological examination of the surgical material was carried out at the National Medical Research Center for Pediatric Hematology, Oncology and Immunology named after Dmitry Rogachev.

Clinical case. A 6-year-old boy fell ill acutely with an increase in body temperature of 38.50 C. During examination in the Central district hospital, a clinical blood test revealed leukocytosis up to 11 thousand / ml with a neutrophil shift, normochromic anemia with hemoglobin 82 g/l, thrombocytosis 874 thousand / ml , accelerated ESR 60 mm/h. Antibacterial therapy was prescribed in a combination of 2 drugs: cefotaxime 610 mg x 3 times intravenously, amikacin 135 mg x 2 times intravenously and heparin, but there was no effect from the treatment. Due to high fever, increasing hyperfibrinogenemia and ESR, with suspicion of multisystem inflammatory syndrome, he was hospitalized at the Pediatric Center of the SAI of the Republic of Sakha (Yakutia) "Republican Hospital No. 1 – NCM named after M.E. Nikolaev".

Upon admission, the child's condition was serious, due to a fever of 39 C, pronounced pallor of the skin was noted,

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peripheral lymph nodes, liver, and spleen were not enlarged. A mass was palpated from under the left hypochondrium +4 cm. Breathing, hemodynamics, and urination were not impaired. A clinical blood test revealed severe normochromic anemia (HB 60 g/l), leukocytosis 11 thousand/ml with a neutrophil shift, thrombocytosis up to 1000 thousand/ml, accelerated ESR 78 mm/h, hyperfibrinogenemia (12 g/l), increased level of D dimer (3.69 μg/ml) and CRP (236 mg/l). Biochemical blood test and general urinalysis showed no pathology.

For diagnostic purposes, computed tomography of the chest and abdominal cavity was performed. An abdominal mass measuring 3*4.5*7.5 cm was detected, with accumulation of contrast agent. The formation is adjacent to the tail of the pancreas and intestinal loops (Fig. 1).

In the oncohematology department, studies were carried out to determine the extent of the tumor process; no signs of metastasis were identified.

An MRI examination was also performed (Fig. 2).

According to MRI and CT data, it was not possible to determine the organ affiliation, so a laparoscopic biopsy was planned to determine the histological affiliation and revision of the tumor. During a laparoscopic examination of the abdominal cavity, a tumor-like formation was discovered on the upper floor on the left, intimately adjacent to the body of the stomach along the greater curvature and transverse colon, measuring 12x8.0 cm. A consultation was created intraoperatively, and, taking into account the high vascularization of the tumor and the possibility of complete tumor removal, it was decided to switch to an open approach. After triple treatment, a transverse incision was made on the left - laparotomy. The tumor-like formation is dislocated into the wound (Fig. 3).

During revision, the formation is intimately adjacent to the body of the stomach along the greater curvature and transverse colon. Using Ligash's apparatus, the formation was cut off and removed. A biopsy of the lymph nodes of the gastrocolic ligament No. 1 and mesenteric transverse colic ligament No. 2 was taken. The formation and lymph nodes were sent for histological examination to the National Medical Research Center for Pediatric Hematology, Oncology and Immunology named after Dmitry Rogachev.

On the second day after removal of the formation, normalization of temperature, ESR, and a gradual decrease in the level of fibrinogen and CRP, platelets to normal are noted.

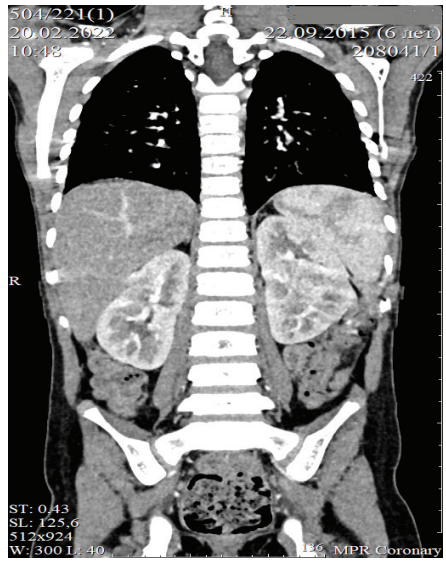

Fig.1. CT scan of the chest and abdomen. Abdominal cavity formation

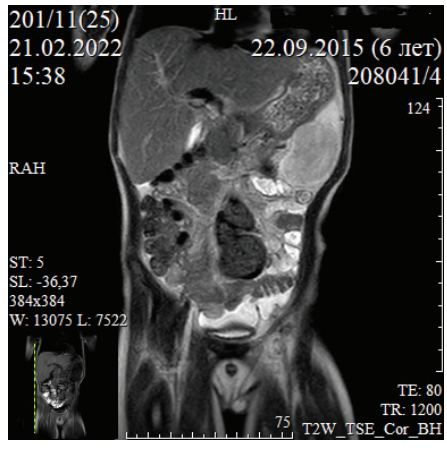

Fig. 2. MRI of the abdominal cavity, formation of the right half of the abdomen

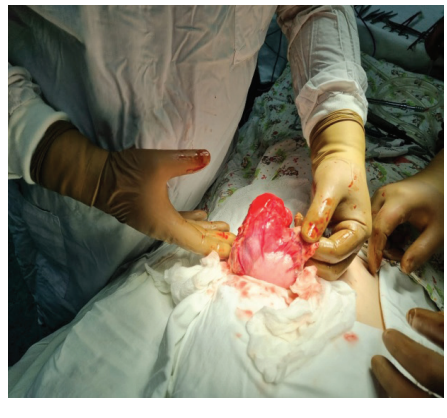

Fig. 3. Photo of the operation: The tumor-like formation is dislocated into the wound

According to histological examination, the tumor is in a thick fibrous capsule, the resection margins are without tumor growth. Neoplastic tissue is represented by long intertwined bundles of medium-sized spindle-shaped cells with a small amount of eosinophilic cytoplasm and ovoid nuclei. The stroma is sclerotic with abundant lymphoplasmacytic infiltration. Expression of SMA, ALKD5F3 was detected. Lymph nodes No. 1, 2 without tumor growth.

The material was further sent for molecular genetic research at the National Medical Research Center of Oncology named after N.N. Petrov". Using reverse transcription PCR and sequencing, unbalanced expression of the ALK gene was revealed, and a chimeric transcript TPM4ex7-ALKex20 was detected. Thus, the child has identified a target for targeted therapy, which can be reserved in case of unresectable relapse of the disease. During the control study, no evidence of tumor recurrence in the child was obtained.

Conclusion. Inflammatory myofibroblastic tumor (IMT) is a rare tumor in children with intermediate biological behavior. The main diagnostic method is histological verification using immunohistochemical methods. The standard of treatment is radical surgical treatment with negative resection margins [4]. If gene expression is present, targeted drugs are used as a second line of therapy or in the case of unresectable tumors.

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