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OSTEOGENIC SARCOMA - CLINICAL CASES IN THE PRACTICE OF A CHILDREN'S ORTHOPEDIST AND SURGEON

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Abstract

Despite the significant achievements of modern medical science, oncological diseases pose great difficulties regarding the etiology, timely diagnosis, treatment and prevention of these ailments. Early diagnosis of tumors is the main task facing doctors. Our clinical observations over the past 5 years have shown that all children with tumors sought medical help late with advanced forms. Therefore, our task is to convey to a wide range of pediatric

specialists, and not only, information that will help to recognize tumors as early as possible. Time is of crucial importance - whether the child will live or not. The given typical clinical cases should alert you to the recognition of tumors. Unfortunately, the markers of osteogenic sarcoma have not been developed at the moment, so clinical signs play an extremely important role in recognizing this disease.

Key words: unossified fibrous bone; pathological fracture; cortical defect; osteochondropathy; features; radiograph; tomography.

Despite the significant achievements of modern medical science, oncological diseases pose great difficulties in terms of etiology, timely diagnosis, treatment and prevention of these ailments [1]. There are dozens of theories of the origin of tumors, but none of them can fully answer the question - how does cancer occur [2, 3]?

In recent years, encouraging steps in this direction have been made by genetics and immunology [4-6]. Thanks to genetic "scissors" (CRISPR/Cas9), in the future it will be possible to cut out those genes that contribute to the emergence of tumors [7]. Genetic correction of the human genome is currently possible for a limited number of hereditary diseases, up to the age of two years of the child and is very expensive (more than three million US dollars), which makes it impossible to use this method for most patients (Nobel Prize Laureates in Chemistry Emmanuel Charpentier and Jennifer Doudney in 2020) [8, 9]. For example, Williams' tumor, which has a dominant type of inheritance [10], may be cured by genetic engineering in the future long before the first clinical manifestations of the disease.

Immunology has made a huge contribution to the treatment of cancer, especially melanoma of the skin and lungs (James Allison and Tasuku Honjo – Nobel Prize winners in the field of medicine in 2018) [11, 12]. If initially immune correction of tumors for children was prohibited, now it has been improved and is widely used. These immune drugs are available in Ukraine at an affordable price [2] .

In the future, these two methods may completely replace such "gold standards" of treatment as chemotherapy, radiotherapy, and surgical interventions. In particular, OS in most cases cannot be treated with radiation therapy. The main emphasis, at the modern level of medicine, is still given to chemotherapy and gentle segmental tumor resections. In some cases, if the OS is large, the entire limb is amputated [13, 14] .

The main task facing doctors is early diagnosis of tumors. Our clinical observations during the last 5 years showed that all children with tumors sought medical help late with advanced forms. Therefore, our task is to convey to a wide range of children's specialists, and

not only, information that will help recognize tumors as early as possible. Time is of the essence - whether the child will live or not. The following typical clinical cases should alert you to tumor recognition. Unfortunately, markers of osteogenic sarcoma have not been developed at the moment [15, 16], so clinical signs play an extremely important role in recognizing this disease. Why is osteogenic sarcoma so important for a doctor? Because it is a rare tumor, it is extremely malignant with high mortality in children and, unfortunately, there is a lack of awareness among doctors who are the first to encounter this tumor. Long-term observations, examinations, incorrect interpretations of auxiliary diagnostic methods and sometimes incorrect treatment steal the "golden time" for helping such a child [16, 21, 22]. Thus, experts from leading clinics in Europe and America claim that with early diagnosis, in some cases, the recovery of a child with OS is possible in 70% of cases. Most OS patients who received adequate treatment live no more than 5-6 years. If the treatment was not carried out, the child "burns out" within a year.

Our most striking cases from practice.

Clinical case 1. Girl N, age 18. While walking, she carelessly hit her knee. There was a slight pain in the leg, which gradually increased, especially at night. The leg was swollen, badly bent in the joint. The local doctor ordered an X-ray examination of the knee joint of the injured leg, but he did not see any changes. She received treatment at home - compresses, ointments, various rubbings. During 2 weeks, the disease progressed. The doctor again ordered an X-ray examination of the knee joint - he did not see any changes. Treatment continued with traditional home remedies - rubbing, compresses, pain pills.

Due to severe pain and poor gait, the girl re-injured her injured knee. She was taken to the district hospital by ambulance. During the examination, they saw pronounced swelling of the knee joint, its pastiness, an increase in the general temperature over 38 C, leukocytosis in the blood, a shift of the leukocyte formula to the left. No changes were seen on the X-ray. Osteomyelitis was suspected. They performed a diagnostic puncture of the injured knee - they received a small amount of liquid that resembled black tar, there was no pus (according to the mother). The leg was fixed with a plaster cast bandage, which the child took off after a day due to unbearable, constant and severe pain, especially at night. The strongest painkillers did not help. In addition, an ultrasound of the knee joint was performed - they could not assess the state of the changes, so they were referred to the regional oncology clinic. During the repeated ultrasound examination, the obtained data were considered as an inflammatory process. At the insistence of her mother, the girl continued treatment in one of the regional institutions. She received anti-inflammatory therapy, vitamins, various physical procedures. The result of the

treatment was that the pain increased, the swelling of the knee increased significantly, the child rapidly lost body weight. On the recommendation of a doctor from one of the private clinics, she was referred for an MRI examination of her leg. The specialists of this institution were not able to assess the changes after the MRI. She was examined several times by a rheumocardiologist, a private surgical doctor. They decided to refer the patient to the Kyiv sports trauma center. Since the results of the previous MRI did not satisfy Kyiv specialists, they recommended a repeat examination in three days (a long queue). Due to severe pain and deterioration of the general condition (exhaustion), the mother refused further examination. They turned to one of the children's medical institutions of the region.

Upon examination, the general condition of the girl is serious. Exhausted, dehydrated, adynamic. Gray skin. The eyes are deeply sunken in the sockets, the nose is pointed. He limps badly on his left leg, the slightest movement causes unbearable pain in his leg. He cannot bend his knee. The lower third of the left thigh is strongly deformed in the form of a large spindle, the skin over the tumor is tense, hot, with a marked venous network (Fig. 1). Peripheral lymph nodes are not enlarged.



Fig. 1. Patient N. Osteogenic sarcoma of the distal metaphase of the left thigh. Flexion contracture of the left knee joint with pronounced pain syndrome. A pronounced increase in the perimeter of the left thigh - "plus tissue". Reactive arthritis of the left knee joint

X-ray images taken several months ago revealed paraosseous tissue growths, a spicule-like periosteal reaction in the thickness of the tissue component on the back surface of the thigh, images of low quality.

On previously performed MRI of the knee joint (Fig. 2), there is diffuse infiltration, swelling of the bone marrow of the distal end of the right femur over the entire length that fell into the field of study (part of the diaphysis and metaphysis) with a length of more than 10 cm in the form of an inhomogeneous hypointense MR signal . Damaged integrity of the cortical layer. Paraaxially, at the height of the metadiaphysis, an asymmetric circular pathological tissue component with a thickness of 0.7-2.4 cm with a spicule-like periostosis that exfoliates the periosteum is determined. The periosteum is locally destroyed on the back surface with the spread of the tissue component into the intercondylar fossa around the cruciate ligaments, and on the front surface above the patella. The muscles of the front group of the thigh are displaced, the upper turn of the kneecap is compressed. A moderate amount of synovial effusion. Signs of chronic damage to the posterior horn of the medial meniscus in the form of a thin transverse strip.

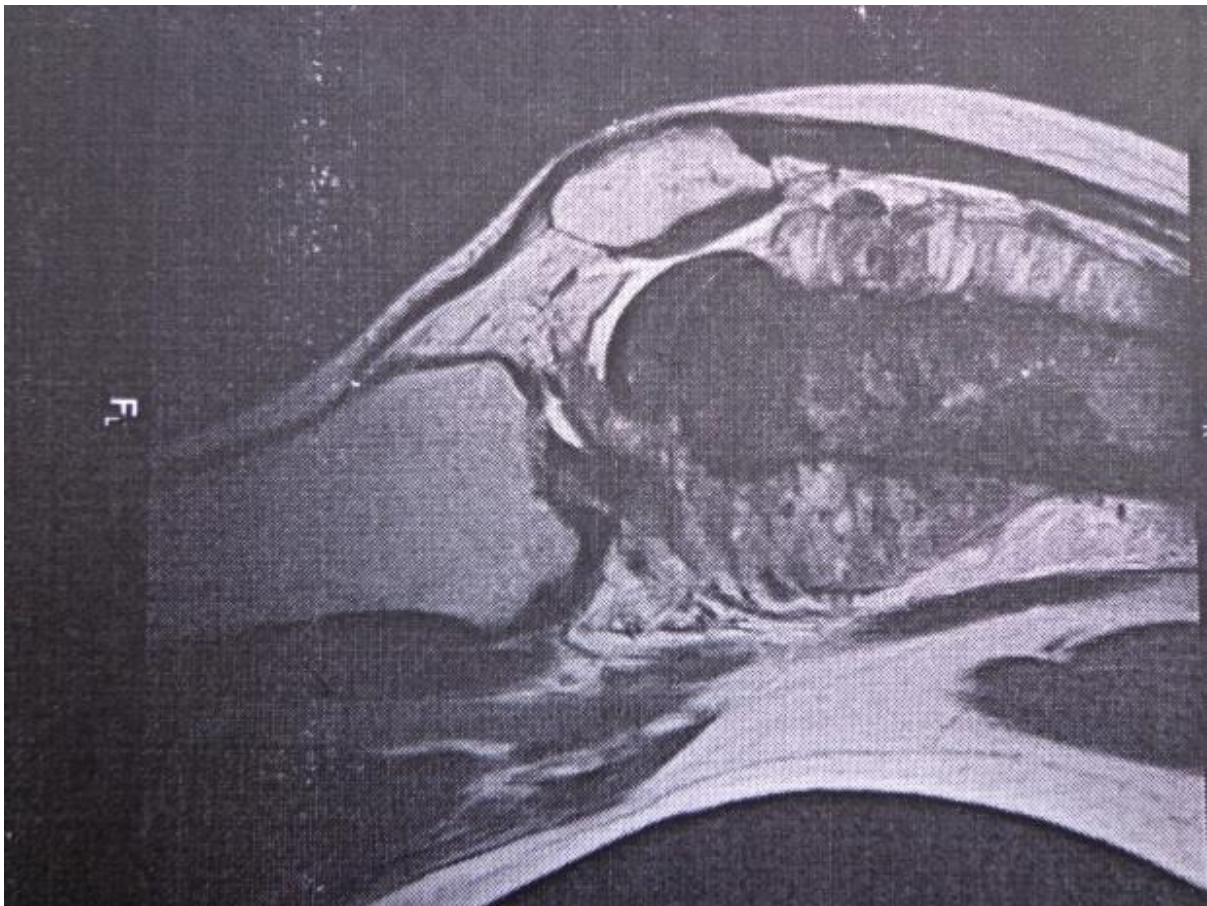


Fig. 2. MRI of the left knee joint of patient N. Pronounced cancerous periostitis, destruction of the distal end of the femur, multiple spicules.

Osteogenic sarcoma of the distal end of the left thigh, osteoblastic component, reactive synovial effusion was diagnosed.

She was immediately referred to the National Cancer Institute in Kyiv. She managed to receive only the initial course of chemotherapy - the child died.

Such a typical clinic of the course of OS was described in 1934 by the classic of purulent surgery Voyno-Yasinetsky in his monograph "Essays of purulent surgery" in a separate section called "Osteomyelitis or sarcoma". Amazing coincidence of these two clinical cases almost 90 years later [17].

Clinical case 2. A 12-year-old girl was hit on the shoulder by a ball. There was swelling, minor pain. She was treated on an outpatient basis for a shoulder injury - ointments, compresses, painkillers, vitamins. The swelling progressively increased and resembled a spindle-shaped thickening. A week after the injury, an x-ray examination of the shoulder was performed - chronic osteomyelitis. The child was sent to the regional clinic. Spindle-like thickening of the proximal end of the right shoulder was revealed, not painful on palpation. The skin is tense, the subcutaneous veins are dilated. Lymph nodes are not enlarged. There are no complaints of pain in the shoulder. The general condition of the child is moderate. Blood parameters are within normal limits. A detailed analysis of the X-ray images obtained showed that the proximal part of the humerus is destroyed, there is cancerous periostitis, multiple spicules, tumor growth into the surrounding tissues (Fig. 3).



Fig. 3. Radiograph of patient M. Osteogenic sarcoma of the proximal metaphase of the right shoulder.

An osteogenic sarcoma of the proximal end of the right shoulder was diagnosed. Referred to the oncology center of the capital for treatment - the diagnosis was confirmed. The further fate of the child is unknown.

In this case, OS was diagnosed "accidentally", the received injury was the trigger for the examination of the child. The extremely fast rate of bone destruction during the week is surprising, which became a big mystery for the doctor who first saw the radiographs and suspected chronic osteomyelitis [17].

Summarizing the results of two clinical examinations, it can be noted that the diagnosis of tumors is the most difficult and responsible part of oncology. The mental potential of the doctor, his experience, intuition, make the diagnostic process creative and correct.

OS most often occurs during puberty (puberty). This is a tumor of a young age, isolated cases occur after the age of 40 [18, 19].

Boys get sick twice as often as girls. Most often, OS nestles in the knee joint (distal metaphase of the thigh, proximal metaphase of the tibia).

The disease begins suddenly, in the midst of complete health . The pain is unbearable, it is poorly anesthetized with analgesics and drugs. The affected limb quickly "swells" [19].

Rapid exhaustion of the child - cancer cachexia, severe intoxication against the background of high temperature. The general blood test is characterized by low hemoglobin values, an increase in the number of leukocytes, rod-shaped neutrophils, which can sometimes be considered as manifestations of acute hematogenous osteomyelitis [17 - 20].

In fact, after three months, 95% of cases have multiple lung metastases (Fig. 4).



Fig. 4. X-ray of the lungs of patient K, age 12, with osteogenic sarcoma of the hip. Multiple metastases in the lungs.

In children, metastases to other bones practically do not occur, the tumor spreads through the arterial network of blood supply.

X-ray examination, which is available in any clinic, is helpful, basic and extremely important. Destruction of the metaphyses of long tubular bones (femur, shoulder), cancerous periostitis, multiple spicules, tumor growth into the surrounding tissues are indisputable signs of the most malignant bone tumor - OS.

Prospects for further research. The world trend regarding OS is the development of oncological markers of this extremely dangerous, deadly disease, especially in children, with the aim of early diagnosis of the tumor. There are great hopes for genetic engineering and immunological correction in children who cannot tolerate chemotherapy, radiation and especially surgical interventions.

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