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A big Wilms tumor in a 14-months-old boy

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Abstract

Introduction. Wilms tumor (nephroblastoma) is a malignant neoplasm located in a kidney. It is typical for children and it occurs almost exclusively in a childhood.

Purpose of work, material and methods. The aim of the study is presentation of symptoms, diagnostics and treatment of the massive Wilms tumor in a 14-months-old child.

Results. In the paper it was presented the case of 14-months-old boy with the Wilms tumor. The patient did not present typical symptoms of a nephroblastoma, except from a perceptible tumor in a abdomen. US and CT scan confirmed a preliminary diagnosis of the neoplasm. After US, a parasitic cyst was taken into account in a differential diagnostics. Despite a growth the tumor after a pre-operative chemotherapy and a radical nephrectomy was successful. A complete remission was achieved.

Discussion. The age of the boy was standard for Wilms tumor. The patient presented the main symptom for nephroblastoma – a large, painless abdominal mass, but without other symptoms (haematuria, malaise and hypertension). A protocol involves a primary treatment with chemotherapy, next surgical resection and later post-operative chemotherapy. The prognosis and survival rate of Wilms tumor depends on pathologic pattern. Relapses are rare in nephroblastoma.

Conclusion. The case was not typical form of the Wilms tumor. Doctors should take it into account while diagnosing other illnesses with similar symptoms.

Key words: Wilms tumor; nephroblastoma; child; treatment.

Introduction

Wilms tumor, also known as nephroblastoma, is a malignant neoplasm located in a kidney. It derives from embryonal kidney precursor cells in which cell growth and differentiation are dysregulated during development [1]. It is associated with specific genetic predisposition syndromes in 10–15% of cases [2]. It is the most common pediatric renal neoplasm [1] with prevalence of one in 10 000 children younger than 15 years of age. It is basically a pre-schooler malignancy with a median age of 3,5 years old [3]. It occurs almost exclusively in childhood [4].

Purpose of work.

The aim of the study is presentation of symptoms, diagnostics and treatment of massive Wilms tumor in a 14-months-old child.

Material and methods.

The case repot is about the 14-months-old boy hospitalized in the Department of Pediatric Hematology, Oncology and Transplantology of Medical University of Lublin due to enlargement of the abdomen. It is a retrospective review of the clinical features of the patient.

Results.

A 14-months-old boy was admitted to the Department of Pediatric Hematology, Oncology and Transplantology due to crucial increased abdominal dimensions on the left side. The patient was in a good general condition without any other symptoms. Past medical, family and social histories were insignificant. A physical examination revealed a big, hard tumor, above the level of the chest, extended from ribs to an anterior superior iliac spine and from posterior axillary line to lateral sternal line on the right side. Blood tests showed elevation of alpa-fetoprotein (AFP): 39,21 ng/ml (reference range: 0-7,9 ng/ml) and also elevated neuron-specific enolase (NSE): 27,03 ng/ml. Abdominal ultrasonography (US) and computed tomography (CT) with intravenous contrast scan visualised a large [81 x 86 x 102 mm (AP x LR x CC)] cystic tumor with septums in the superior pole of the left kidney (Figure 1, 2).

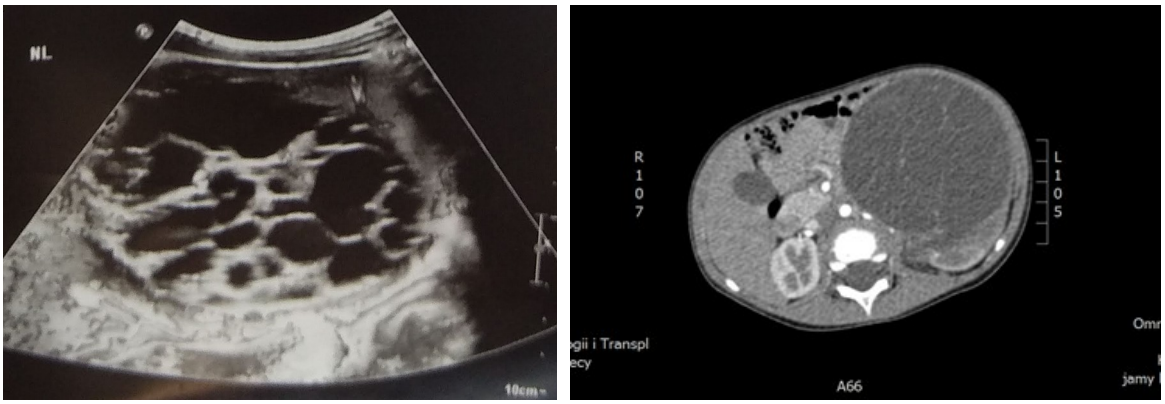

Figure 1, 2 : Before treatment

The Wilms tumor was diagnosed based on the above studies. Metastases were not found in the CT scan of the chest. After one week the patient started a treatment according to European

Programme – SIOP. The boy received pre-operative chemotherapy: alternatively vincristine and vincristine in combination with actinomycin for three weeks. A control ultrasonography and CT of the abdomen also CT of the chest were performed. Tests revealed a tumor progression [dimensions of tumor: 92 x 97 x 119 mm (AP x LR x CC)], without metastases in the chest (Figure 3).

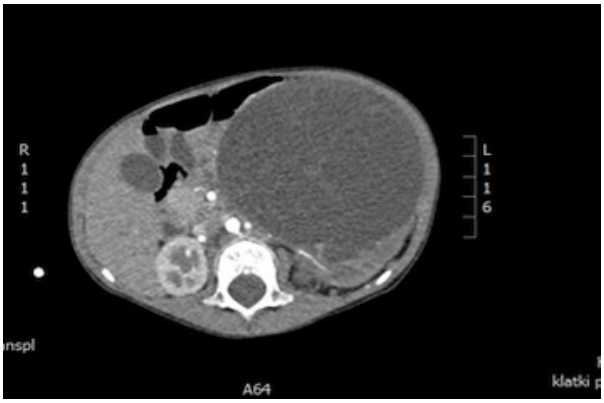

Figure 3: After chemotherapy

The patient got the vincristine. Due to a delayed surgery, the patient received one additional dose of vincristine and it ended a pre-operative chemotherapy. One week later the tumor was resected - a radical left-sided nephrectomy. Cystic partially differentiated nephroblastoma was identified in a histophatology. The boy received a post-operative chemotherapy: alternatively vincristine and vincristine in combination with actinomycin and it ended the treatment. After two weeks a control US and CT scan were performed. Tests showed no recurrence, what was a sign of complete remission (Figure 4).

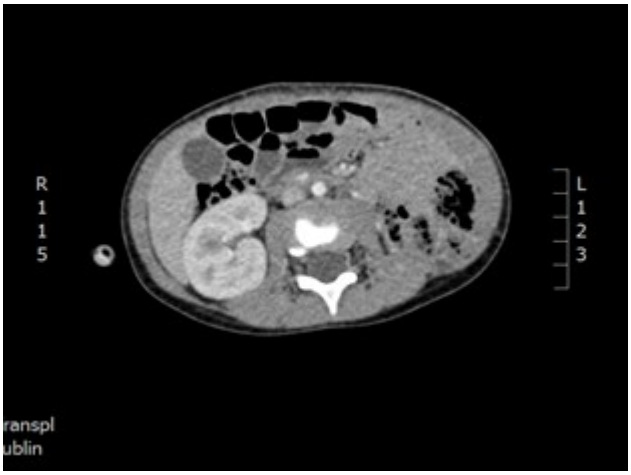

Figure 4: After treatment

Discussion.

The age of the patient in the present study was 14 months, which was consistent with the age of onset reported in the literature [3,4]. Wilms tumor usually manifests as a large, painless abdominal mass and more less as a haematuria, pain, malaise and hypertension [5, 6]. Up to 10% of nephroblastoma is discovered incidentally after trauma, 25% have microscopic haematuria and 25% manifest as a secondary hypertension related to renin production [6]. In this study the physical examination revealed the abdominal tumor, but the patient did not present other symptoms. The primary distant site for metastases are lungs [5], so the patient was subjected to CT scan control and it did not show metastases. Generally imaging of Wilms tumor begins with USG which may evaluate whether the mass is intrarenal or extrarenal and whether it is solid or cystic. It should be noted that often Wilms lesions appear to have a large hypoechoic areas due to central necrosis and cyst formation. Hyperechoic areas can represent areas of fat, calcification or hemorrhage. It may also appear less commonly as a solid spherical mass [6]. In this case the mass was extrarenal, with hypoechoic areas. A parasitic cyst was taken into account in a differential diagnostics. Next CT scan is recommended in order to better stage and anatomically define the primary tumor and assess for distant metastases [5]. Diagnosis of nephroblastoma requires histological confirmation [7]. In the study it is a "cystic partially differentiated nephroblastoma". Staging is determined by the anatomic extent of the tumor. Most patients with Wilms tumor can be cured. Protocol involves primary treatment with chemotherapy to decrease the risk of intraoperative rupture and hemorrhage and to reduce the disease stage at the time of surgical resection and later post-operative chemotherapy [7]. In this case the boy received prolonged pre-operative chemotherapy due to delayed surgery. Despite the pre-operative chemotherapy, the tumor mass increased. Initially it was considered an unsuccessful prognosis, but it was possible to remove the tumor while the surgery. After completed protocol of treatment, it turned out, that it was possible to achieve a complete remission. The prognosis and survival rate of Wilms tumor depends on pathologic pattern, age at the time of diagnosis and extent of disease [8], but nephroblastoma is one of the few malignancies which have a better overall survival despite being diagnosed at advanced stages [3]. Relapses are rare in Wilms tumor [8]. Four-year relapse-free survival is 91% for stage I, 88% for stage II, 79% for stage III, and 78–84% for stage IV [9]. In this study it is too early to rate four-year relapse-free survival, because the patient is only six month after a termination of treatment.

Conclusions.

Presented case is an unusual example of Wilms tumor, because the patient did not present classical symptoms of a nephroblastoma, except from enlargement of abdominal dimensions. Parents should pay attention to an untypical changes in the child. An initial progression of tumor growth (despite the chemotherapy) does not decide about bad prognosis.

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