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Massive bilateral Wilms tumor and an effective therapy – case report

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Abstract

Introduction. Wilms' tumor is one of the most common cancer arises into an abdomen and it is estimated to be over 90% of all pediatric kidney tumors. The tumor usually arises in a single kidney.

Aim of the work. The aim of our study was to present diagnostics and treatment of massive bilateral Wilms' tumor in a child with abdominal enlargement without any other symptoms.

Material and methods. The case repot is about the 10-months-old boy hospitalized due to enlargement of the abdomen. All diagnostics and treatment was managed by University Children's Hospital in Lublin.

Case report. A 10-years-old boy was admitted to the hospital due to rapid abdominal enlargement. CT scan revealed massive bilateral tumor derived from kidneys. Considering clinical picture and additional tests it was decided to recognize the bilateral Wilms' tumor and implemented chemotherapy according to SIOP 2001 and changed to Umbrella protocol because of inefficiency. A right partial and left nephrectomy was conducted. Post-surgical chemotherapy was applied and finished without complications. There is no recurrence of disease. Kidney functioning is good.

Discussion. The case is asymptomatic abdominal enlargement what is common in patients with Wilms' tumor. Abdominal pain is the most common initial presenting symptom. Picture diagnostic as a CT and US, gives first tip for diagnosis. Clinical picture and CT scans allow to apply chemotherapy. Surgery is essential in the treatment. In bilateral disease kidney function is essential. NSS is recommended.

Conclusions. The only symptom the of the huge tumor was enlargement of the abdomen. In a few cases Wilms's tumor is bilateral and not responding to standard chemotherapy. Partial nephrectomy can provide complete remission and good kidney function.

Key words: bilateral Wilms' tumor; inefficient chemotherapy; partial nephrectomy; complete remission

Introduction

Wilms' tumor (nephroblastoma) is the second most common cancer arises into abdomen. This embryonal type of renal cancer is fifth of neoplasms in children, what is 6% in this age group. Wilms' tumor is estimated to be over 90% of all pediatric kidney tumors. Children before five years old are the major burdened group. There are 75% of nephroblastoma cases. Wilms' tumor can concerns adolescents or adults, but this is only less than 1% of all renal tumors. The tumor usually arises in a single kidney [1, 2]. Bilateral disease is presented in about 5-7% patients. It can develop either synchronously or metachronous. Treatment management of child with bilateral Wilms' tumor is very demanding, because of strong efforts of maintaining as much renal parenchyma as possible to prevent renal failure [3]. The treatment involves pre-operative chemotherapy and surgery which is critical for recuperation [1].

Aim of the work

The aim of our study was to present diagnostics and treatment of massive bilateral Wilms' tumor in a child with abdominal enlargement without any other symptoms.

Material and methods

The case repot is about the 10-months-old boy hospitalized in the Department of Pediatric Hematology, Oncology and Transplantology of Medical University of Lublin due to enlargement of the abdomen. The patient was diagnosed by the medical history, physical examination, laboratory imaging tests and histological examination. The entire treatment was managed by above department.

Case report

A 10-months-old boy was admitted to the Department of Pediatric Hematology, Oncology and Transplantology of Medical University of Lublin for carrying out diagnostics and treatment. From the parents' report, within last 2 weeks they noticed enlargement of the abdominal girth and hardness. General condition of patient during admission to the hospital was good, he seemed not to have pain.

In physical examination a large, hard and fix tumor was found in the left part of the abdominal cavity. The tumor reached from the rib arch to the pelvis, crossed the midline of the body. In laboratory test only elevation of thrombocytes and NSE levels were observed. Ultrasonography (US) showed two tuberous solid changes in both kidneys; left 92x80 mm, right 53x50 mm. Computed tomography (CT) scan revealed (Fig. 1, 2) nodular changes in both kidneys with heterogeneous density (~40 HU); right kidney – 53x63x50mm (RLxAPxCC); left kidney – 89x94x95 mm (RLxAPxCC), which crossed the midline of the body and moved the intestinal loops. In the upper pole of the left kidney was another nodular change: 36x24x41 mm (APxRLxCC). The widening the left pelvis system was observed too. There were no metastases, pulmonary included.

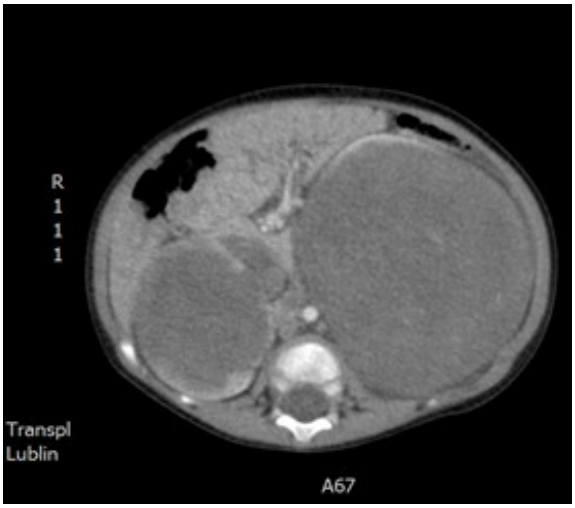

Figure 1: During diagnostics, before treatment

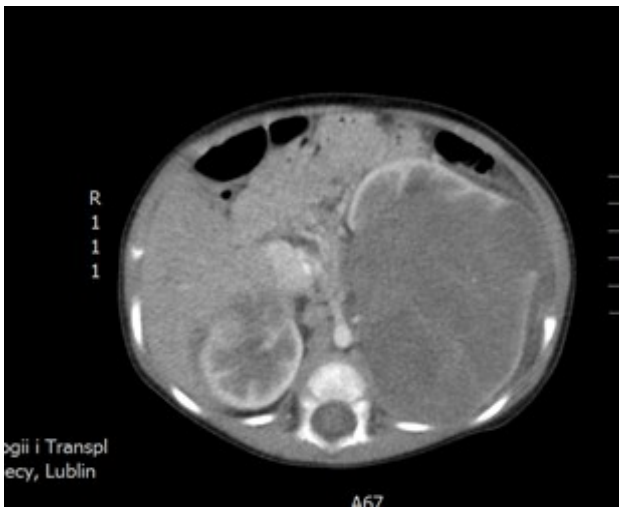

Figure 2: During diagnostics, before treatment

Considering clinical picture and additional tests it was decided to recognize the bilateral Wilms' tumor and began treatment according to SIOP 2001 protocol (VCR, ACT-D). During chemotherapy, a double rise of the temperature to 38^{O}C occurred, without

complications. After 4 blocks of the chemotherapy control tests were carried out, which revealed small reduction in tumors mass. It was made a decision to change chemotherapy to VP/CAR blocks according to Umbrella protocol with good tolerance of treatment. After 2 blocks, preoperative treatment was finished. CT scan revealed (Fig. 3, 4) disease progression: the left kidney tumor – 70x78x61mm (RLxAPxCC), right kidney tumors:11x98x107 mm (RLxAPxCC), upper pole: 60x54x44 mm (RLxAPxCC). A bilateral widening pelvis system was observed.

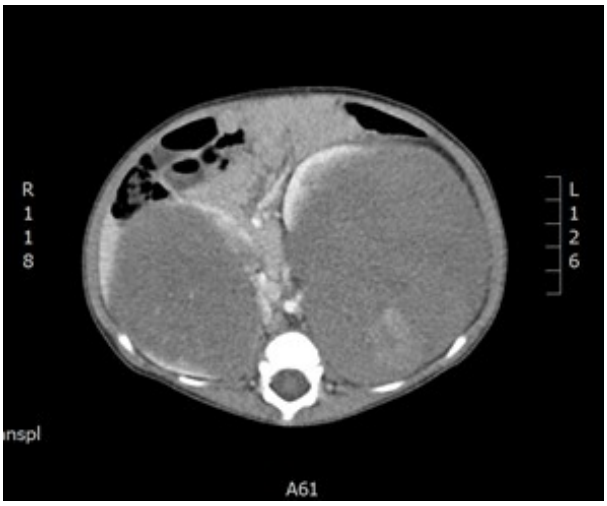

Figure 3: After presurgical chemotherapy

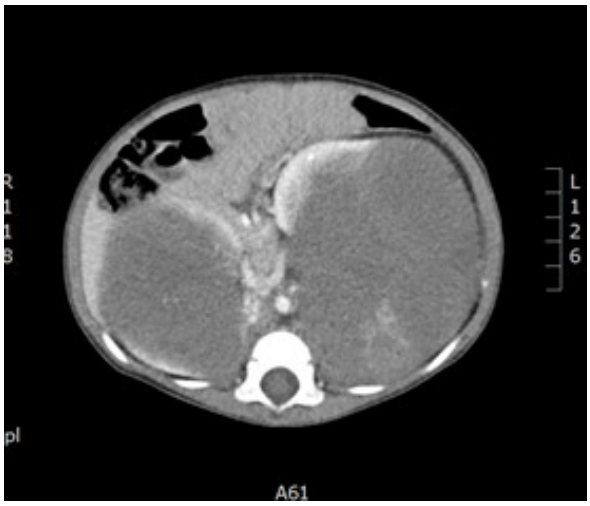

Figure 4: After presurgical chemotherapy

After 3 months from the diagnosis a surgical procedure was performed– partial nephrectomy of the right kidney with tumor removal. A month later it was performed a left nephrectomy with removal of left adrenal gland and surrounding nodes. The histopathological examination confirmed the diagnosis – Wilms' tumor, stroma rich, 10% necrosis without metastases in the nearby tissues and nodes. The VCR/ACT-D postsurgical chemotherapy (SIOP 2001, stage II) was implemented considering an overall clinical picture and finished after 9 months from the diagnosis, without progression of disease in MRI (Fig. 5, 6). Now the disease is stable, there are not any signs of recurrence. Laboratory test presenting a good functioning of the kidney.

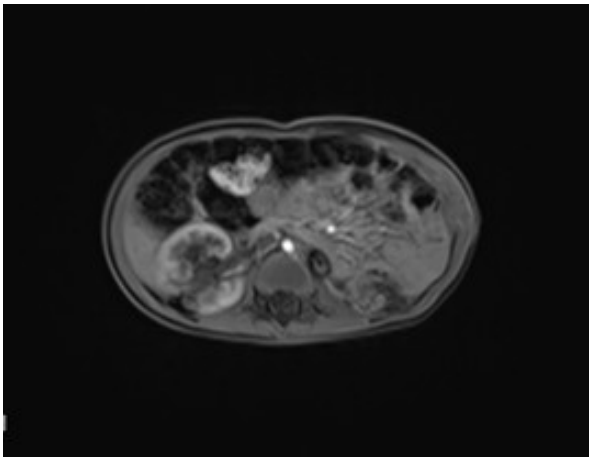

Figure 5: After the surgery

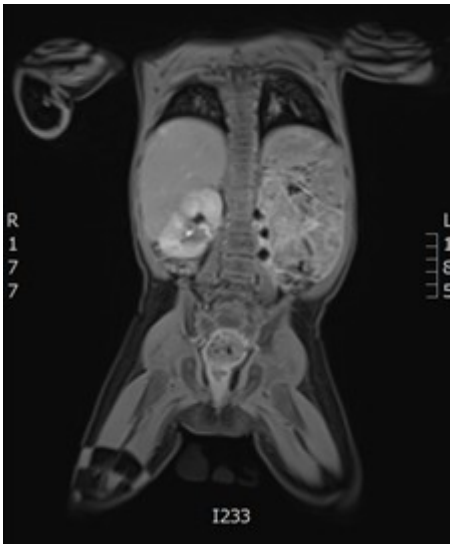

Figure 6: After the surgery

Discussion

Most of the Wilms' tumors reveal as a asymptomatic mass located in an abdomen. There are usually discovered by parents who notice an enlargement of the child's abdomen. In approximately 20-30% of cases presenting symptoms as a: abdominal pain, malaise, microscopic or macroscopic hematuria, urinary tract infections, hypertension, fever etc. Abdominal pain is the most common initial presenting symptom (30-40%). Another are hypertension (25%) and hematuria (12-25%) [1, 4]. In our case there were not any symptoms of disease in child. The only feature that draws attention was an enlargement of the abdomen noticed by parents in a short period of time – 2 weeks. Laboratory tests did not reveal deviations from the norm, the most common in Wilms's tumor included. First tip for diagnosis was obtained by using picture diagnosis: US and CT, what showed the kidneys' origin of the tumor. Imaging and diagnosis of Wilms tumor generally begins with US. It provides information if the change is solid or cystic, intra- or extra-renal. Wilms tumor lesions appear to have large hypoechoic areas due to central necrosis and cyst formation. US and CT are useful to assess patency of vessels what is important in 5-10% cases with vascular invasion. MRI and CT are imaging modalities, which are vital in diagnostics, but the MRI is now preferred. Both are able to visualize a normal renal tissue and tumor. In a bilateral tumor MRI is superior to CT and helps with renal preservation surgery [5].

Imaging modalities made in the clinic showed changes typical for a Wilms tumor. No metastases were revealed in CT. Pulmonary metastases are the most common in children with

Wilms tumor. There are present in 10-20% cases at the time of diagnosis. Treatment includes pre-operative chemotherapy and surgery according to SIOP protocol. It consists of double-agent chemotherapy (Vinciristine and Actinomycin D) in children with bilateral tumors [1, 6, 7]. The patient had localized disease so SIOP protocol was implemented. In control tests the tumor turned out to be nonresponsive. In instances of tumor nonresponsiveness switching to treatment with etopside and carboplatin is recommended. After preoperative chemotherapy, radical tumor nephrectomy is the standard of care for children with Wilms tumor. In bilateral disease, in which kidney function is essential, a nephron sparing surgery (NSS) is considerably better than when over types of surgery is used. NSS is now acceptable for nonsyndromic unilateral Wilms tumors under certain conditions: small tumor volume (<300 ml) and the expectation of a substantial remnant kidney function in patients without nodes' metastases [7]. Left kidney tumor was too large to proceed partial nephrectomy, in contrast to change in right kidney where that procedure was performed. According to SIOP protocol, stage II, post-operative chemotherapy was implemented [3, 8]. The preservation of good longterm renal function after treatment is important. Nevertheless, the surviving individual may develop renal failure or late effects due to anticancer treatment. The incidence of end-stage renal failure for bilateral tumor was 11,5%, according to the sources [3].

Conclusions

The presented case is an example of a cancer which the only symptom was enlargement of the abdomen. Considering the Wilms' tumor biology, the rapid change in the abdominal circumstance should be an alarm symptom for the child's parents. In a few cases Wilms's tumor is bilateral and not responding to standard chemotherapy. Surgery is essential in the treatment of Wilms' tumor. Even a bilateral and very massive tumor can be treated with good results, without significant kidney failure. Immediate implementation of treatment is very important in the prognosis.

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