Kwolczak Justyna, Roszkowska Anna, Winiarz Aleksandra, Świerszcz Łukasz, Winiarz Marek, Nurzyńska-Flak Joanna, Piecewicz-Szczęsna Halina. Relapsed Wilms' tumor with pulmonary metastases – a case report. Journal of Education, Health and Sport. 2018;8(9):1629-1634. eISNN 2391-8306. DOI http://dx.doi.org/10.5281/zenodo.1438661
http://ojs.ukw.edu.pl/index.php/johs/article/view/6130

The journal has had 7 points in Ministry of Science and Higher Education parametric evaluation. Part B item 1223 (26/01/2017).
1223 Journal of Education, Health and Sport eISSN 2391-8306 7

The authors declare that there is no conflict of interests regarding the publication of this paper.
Received: 14.08.2018. Revised: 15.09.2018. Accepted: 15.09.2018.

Relapsed Wilms' tumor with pulmonary metastases – a case report

Justyna Kwolczak[1], Anna Roszkowska[1], Aleksandra Winiarz[2], Łukasz Świerszcz[1], Marek Winiarz[1], Joanna Nurzyńska-Flak[3], Halina Piecewicz-Szczęsna[4]

Corresponding author: Justyna Kwolczak, orcid.org/0000-0002-3260-5165,
e-mail: j.kwolczak@gmail.com

[1] Student Scientific Circle at the Department of Pediatric Hematology, Oncology and Transplantology, Medical University of Lublin

[2] Student Scientific Circle at the Department of Epidemiology and Clinical Research Methodology, Medical University of Lublin

[3] Department of Pediatric Hematology, Oncology and Transplantology, Medical University of Lublin

[4] Department of Epidemiology and Clinical Research Methodology, Medical University of Lublin

Abstract

Introduction: Wilms' tumor is the most common malignant renal tumor in childhood. It accounts for about 95% of kidney tumors diagnosed in children. The delay in diagnosing neoplastic disease in children at an early stage is due to the occurrence of asymptomatic mass in abdomen as the only sign of the disease.

Case report: We discuss a 5-year-old boy case, with Wilms' tumor of the right kidney. Diagnosis was based on the results of imaging examinations; no fine needle aspiration biopsy was performed. The patient underwent nephrectomy and preoperative and postoperative chemotherapy. During the follow-up period, 11 months after the end of treatment, the patient developed respiratory symptoms. Histopathological examination confirmed tumor metastases of Wilms in the lungs.

Discussion: Oncological vigilance comprises challenge for contemporary medicine. It is necessary to take into account the oncological status of the patient regardless the time of the last check-up in the oncological clinic, especially due to the fact that childhood cancers are characterized by dynamic growth.

Key words: pediatric oncology, Wilms' tumor, oncological vigilance

Introduction

Pediatric solid tumors make up 50% of all pediatric cancers. Wilms' tumor (nephroblastoma) is one of the most common type of solid tumor found in children accounting for up to 6% of all childhood malignancies (1). The total number of new cases in Poland reaches 60 -70 per year. The vast majority of patients is aged from 1 to 5 years old at the time of diagnosis and the peak age of the disease is 3.5 years old (2). There is no significant difference in terms of gender. Bilaterality occurs in 4% to 7 % of cases and tend to present at an earlier age (3). Wilms' tumor is usually sporadic but 1% of all cases is said to be familial and is transmitted in an autosomal dominant way. In about 10% of cases, the Wilms tumor is part of the genetically determined disease syndromes - Beckwith-Wiedemann syndrome, Denys-Drash syndrome, WAGR syndrome, Perlman syndrome or coexists with other congenital malformations - urinary tract defects, aniridia, partial body hypertrophy or skeletal system defects. Wilms 'tumor is very rarely diagnosed in adolescents and adults, according to the study by Mitra et al. the incidence of Wilms' tumor in patients above sixteen years of age is 0.19 / million / year.

Most cases (80–90%) of Wilms tumor present with an asymptomatic abdominal mass. For this reason, a large proportion of patients are admitted to the clinic at an advanced stage of the disease when the tumors cross the midline of the body, emphasizing the abdominal wall (4). Sometimes there are discreet and uncharacteristic clinical symptoms that may suggest other illnesses include: abdominal pain (30 to 40 percent of patients), microscopic or gross hematuria (12 to 25 percent), fever, hypertension or hypotension, loss of appetite, nausea , vomiting, malaise, urinary tract infection, varicocele. If the patient has metastases to lungs, shortness of breath or rapid breathing may occur as one of the symptoms. Usually, the child is in good general condition, symptoms of cachexia are rarely observed. Particularly disturbing symptoms in a child with Wilms' tumor are sudden abdominal pain, anemia, tachycardia and fever (5). Common occurrence of the above symptoms may suggest the occurrence of perinephric or subcapsular hemorrhage.

Most patients with Wilms tumor can be treated by means of multidirectional combination therapy including chemotherapy, surgery or/and radiation therapy in sequences

depending on the stage of the disease and histopathological classification. Multidisciplinary treatment of Wilms tumor led to a significant improvement in overall survival reaching up to 90% for limited disease cases and 75% for metastatic tumors (6).

Case report

Parents and a five-year old boy reported to the Emergency Department because of sudden, acute pain in abdomen. Basing on imaging tests Wilms' tumor was detected. The patient underwent pre-treatment chemotherapy for 4 weeks. CT scan suggested spontaneous Wilms tumor rupture, however, during the surgery it was considered that the neoplastic lesion was completely removed together with the intact capsule. It appeared challenging for the therapeutic team to decide on the use of adequate chemotherapy in relation to the clinical stage of the disease. Due to the ambiguous clinical picture of the tumor, it was decided to use therapy as for a third grade tumor owing to the risk of tumor cell dissemination into the abdominal cavity. During the follow-up visits, the patient did not report any ailments. Nevertheless, 11 months after the end of intensive post-operative chemotherapy - the boy had a fever, presented increased respiratory effort and excessive sweating. 2 - weeks before the occurrence of the mentioned symptoms the boy spent active holidays in the mountains. Basing on the clinical picture and physical examination general practitioner implemented antibiotic therapy. Deteriorating condition of the child alarmed parents who reported to the Emergency Department. X-ray of the chest revealed fluid in the left pulmonary cavity. The patient was admitted to the Department of Pediatric Surgery to drain the pleural cavity, but little amount of the drained fluid and lack of improvement suggested seeking further examination. CT scan showed a solid tumor within the left lung. Unfortunatelly, pathological mass had spreaded to the pleural. Histopathological examination confirmed the relapse of Wilms' tumor. Currently, patient is preparing to the radiotherapy.

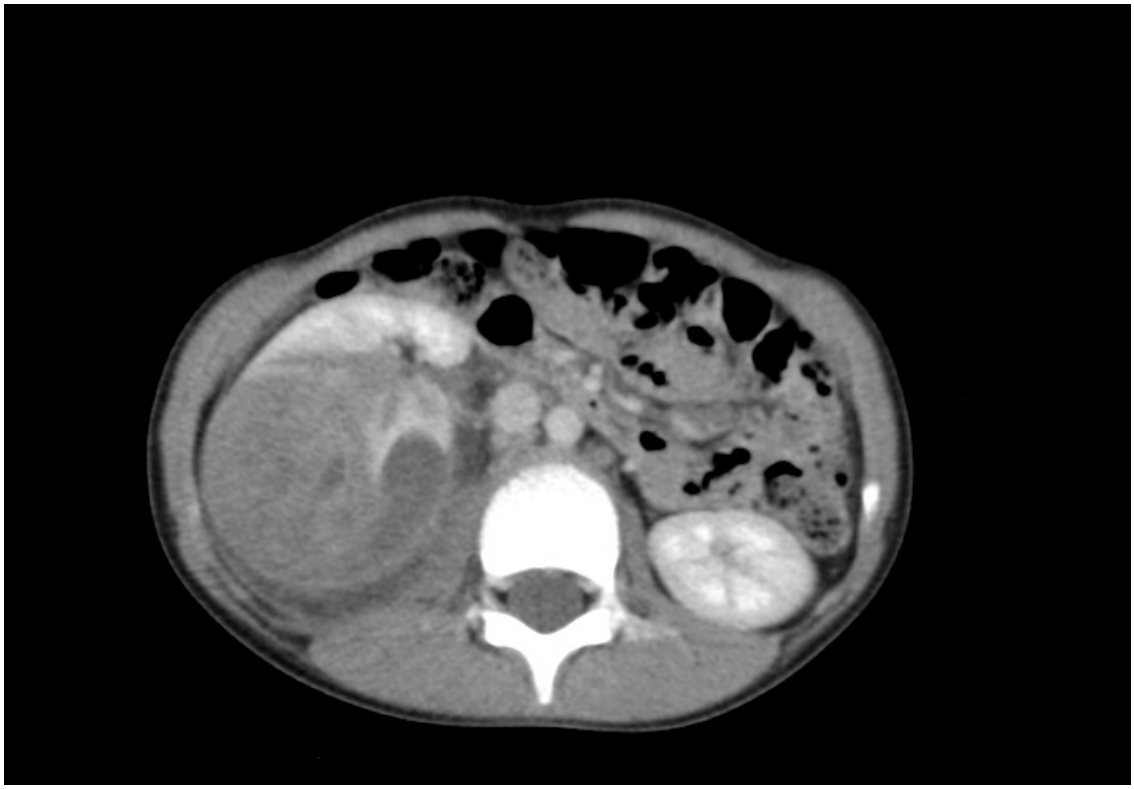

Figure 1. Wilms' tumor of the right kidney.

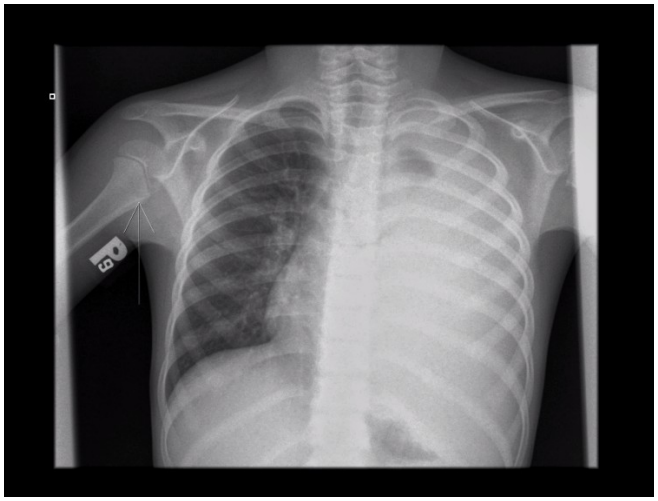

Figure 2. Pleural effusion in the left lung.

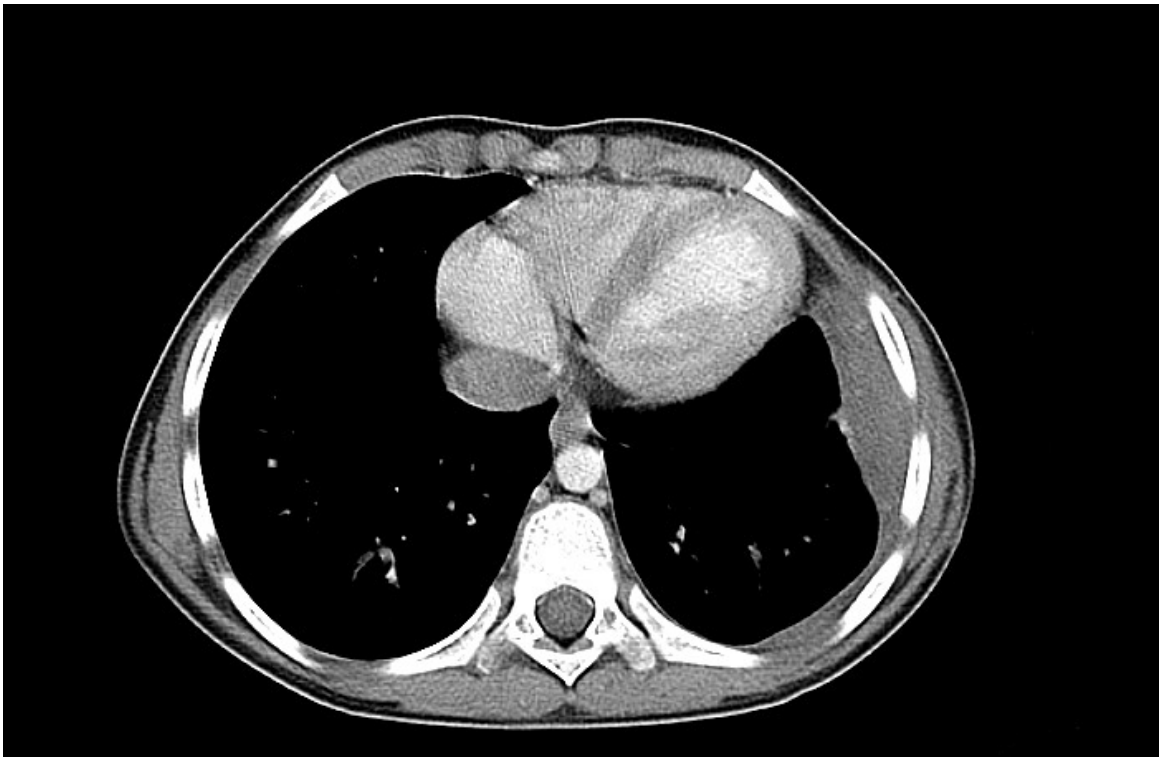

Figure 3. Metastases infiltrating pleura.

Disscusion

Wilms tumor can grow for a long time without giving any characteristic clinical symptoms. Therefore, it is often detected accidentally by parents who sensed a pathological mass in the abdominal cavity, for example during a child's bath (4). In this case, the first symptom was a sudden, strong stomach ache referred by the child. The performed imaging studies revealed nodular formation within the right kidney. However, the overall picture of the change was ambiguous – it suggested tumor bag rupture, but there was no definitive confirmation of such state. Due to the stable condition of the patient, it was decided to postpone the surgical treatment and the use of 4-week preoperative chemotherapy as the first line treatment.

Treatment of fetal nephroblastoma in children is of complex nature. In European centers cooperating within the Inernational Society of Pediatric Oncology (SIOP), it is recommended to administer prechemiotherapy for all patients over 6 months of age, lasting 4 or 6 weeks. The duration of preoperative chemotherapy depends on the stage of the disease, which is determined on the basis of the clinical picture and imaging result, without histopathological verification (7).

The surgery should be performed during the last week of 5-week treatment. Radical nephrectomy is the method of choice for treatment of unilateral Wilms tumor when the second kidney is healthy. During the surgery, material is collected and then pathomorphological examination is performed in order to determine the clinical and pathological advancement, to precisely consider stage of the disease. According to the stage of the disease, the type and duration of postoperative chemotherapy is determined, which in some cases is supported by radiotherapy.

In the United States, the treatment is carried out in accordance with the guidelines of the National Wilms' Study Group (NWSG), which recommends performing an operation as the first line of treatment, without the use of pre-operative chemotherapy.

According to the Polish Pediatric Group for the Treatment of Solid Tumors, the distribution of the stages of Wilms tumor progression in Polish children is as follows: stage I (1.4%), stage II (28.6%), stage III (61.4%), stage IV (7.1%), stage V (1.4%). At present, 85% of patients with fetal nephroblastoma can be cured. However, 15% of children have recurrent Wilms tumor (8). Recurrence of Wilms tumor occurs in the lungs (58%), abdominal cavity (29%) and other less frequent locations (bones, brain, mediastinum). The lung tumor may spread to the pleura, in which case the first manifestation of the metastases will be pleural effusion. Most relapses (90%) appear within the first four years after treatment. For this reason, the oncological alertness of physicians is of great importance in capturing the first alarming symptoms of cancer recurrence.

References:

1. Provenzi VO, Rosa RFM, Rosa RCM, Roehe AV, dos Santos PPA, Faulhaber FRS, i in. Tumor size and prognosis in patients with Wilms tumor. Rev Paul Pediatr. marzec 2015;33(1):82–7.

2. Zatonski M. Epidemiologia Nowotworów Złośliwych w Polsce [Malignant Cancer Epidemiology in Poland]. Potrykowska Strzelecki Z Szymborski J Witkowski J Eds Zachorowalność Umieralność Na Nowotw Sytuacja Demogr Pol Cancer Incid Mortal Demogr Situat Pol; Dostępne na: http://www.academia.edu/9186528/Epidemiologia_Nowotwor%C3%B3w_Z%C5%82o%C5%9Bliwych_w_Polsce_Malignant_Cancer_Epidemiology_in_Poland_

3. Millar AJW, Cox S, Davidson A. Management of bilateral Wilms tumours. Pediatr Surg Int. lipiec 2017;33(7):737–45.

4. Davidoff AM. Wilms Tumor. Adv Pediatr. 2012;59(1):247–67.

5. Byerly D, Coley B, Ruymann F. Perirenal hemorrhage as first presentation of Wilms tumor. Pediatr Radiol. 1 lipiec 2006;36(7):714–7.

6. Szychot E, Apps J, Pritchard-Jones K. Wilms' tumor: biology, diagnosis and treatment. Transl Pediatr. styczeń 2014;3(1):12–24.

7. Bhatnagar S. Management of Wilms' tumor: NWTS vs SIOP. J Indian Assoc Pediatr Surg. 2009;14(1):6–14.

8. Brok J, Lopez-Yurda M, Tinteren HV, Treger TD, Furtwängler R, Graf N, i in. Relapse of Wilms' tumour and detection methods: a retrospective analysis of the 2001 Renal Tumour Study Group–International Society of Paediatric Oncology Wilms' tumour protocol database. Lancet Oncol. 1 sierpień 2018;19(8):1072–81.