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CASE REPORT

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Bilateral nephroblastoma – case report

Elżbieta Łuczyńska[1], Joanna Anioł[1], Andrzej Stelmach[2], Janusz Jaszczyński[2], Andrzej Strach[2]

[1] Zakład Radiologii i Diagnostyki Obrazowej, Centrum Onkologii im, Marii Curie-Skłodowskiej, Oddział Kraków, Kraków, Polska
[2] Klinika Chirurgii Onkologicznej, Centrum Onkologii im, Marii Curie-Skłodowskiej, Oddział Kraków, Kraków, Polska

Author's address: Elżbieta Łuczyńska, Zakład Radiologii i Diagnostyki Obrazowej, Centrum Onkologii im. Marii Curie-Skłodowskiej, Oddział Kraków, Kraków, Polska, e-mail: dgn.ela@interia.pl

Summary

Background: Wilms tumor is the most common renal tumor in children Synchronous bilateral Wilms tumor (BWT) accounts for 5% of all patients registered to the National Wilms Tumor Study Group (NWSTG),

Case Report: A 28-year-old female patient was presented to Oncology Institute with right kidney tumor. Her left kidney was resected due to Wilms tumor in the sixth month of her life.

Abdominal ultrasound was performed and demonstrated a massive right kidney tumor.

Then the abdominal CT was undertaken with the use of 16-slice CT scanner, revealing rotation displacement of the right kidney, with signs of compensative overgrowth and displaced upwards. In the lower and peripheral part of the kidney a nodular mass 7×10×9 cm in size was visible. The tumor was well-demarcated, showing heterogeneous contrast enhancement, extending from the inferio-lateral renal pole. The tumor was adjacent to iliopsoas muscle and abdominal cavity walls; no infiltration of those structures was noted. Described lesion surrounded inferio-medial part of the kidney, approaching kidney pelvis without infiltrating them. Supero-anterior and medial part of the kidney showed normal structure with correct contrasted urine excretion. Lymph nodes enlargement within periaortal area was not detected. Surgical procedure was performed resulting in excision of the tumor with kidney preservation. Histopathology examination revealed nephroblastoma. The tumor was surrounded by a thin, fibrous capsule. Surrounding parenchyma and far tissue waere not involved. The CT examination performed 4 months after nephron sparing surgery revealed: right kidney of 137×51 mm in size, normally located, with correct structure and function.

Conclusions: Multislice abdominal CT have clearly visualized the tumor of the single kidney, showing precise localization of the mass with relation to the kidney pelvis and vessels, allowing for nephron sparing surgical excision.

Key words: Wilms tumor • multislice CT

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Background

Wilms' tumor is the most common renal tumor in children. There are two main cooperative groups dealing with this pathology in the world: The National Wilms' Tumor Study Group (NTWS) and International Society of Pediatric Oncology (SIOP). These groups approaches to therapy [1].

The North American approach (NATS) is concentrated on initial nephrectomy followed by adjuvant therapy, whereas the European approach (SIOP) is based on initial imaging followed by chemotherapy before surgical intervention. The SIOP experience is based on the observation, that this approach gvarantees fewer intraoperative ruptures.

Synchronous bilateral Wilms' tumor (BWT) accounts for 5% of all patients registered to the National Wilms Tumor Study Group (NWSTG), with only few patients treated at a single institution.

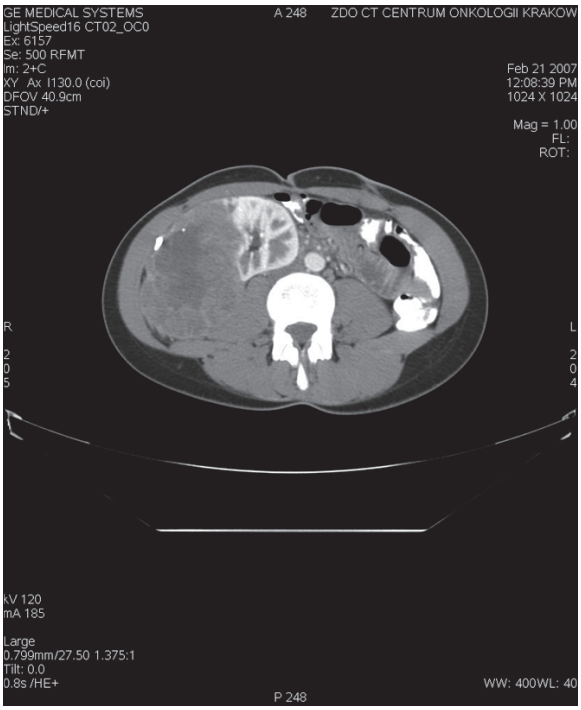

Figure 1. Abdominal CT – post contrast phase. Axial image. Irregular tumor of the right kidney.

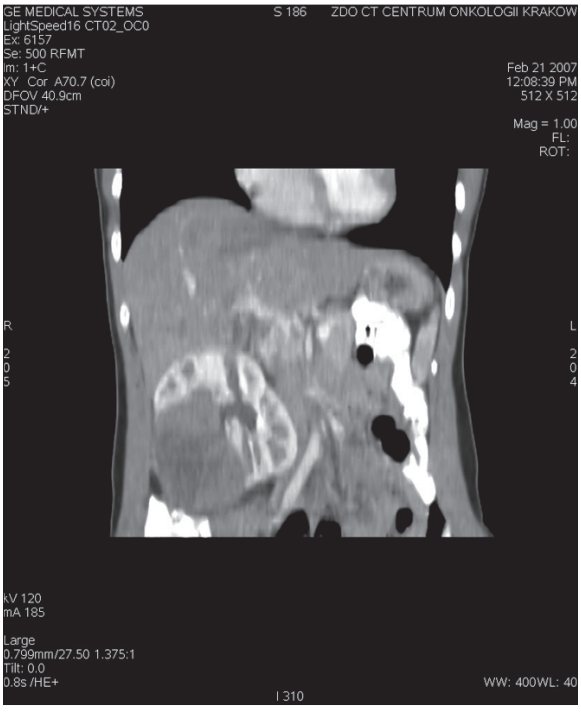

Figure 2. Abdominal CT – post contrast phase. Coronal reconstruction. Irregular tumor of the right kidney.

The accuracy of diagnostic imaging methods especially computed tomography (CT) to determine the extent of tumor involvement has been evaluated.

The importance in the preoperative staging results from the possible role of preoperative chemotherapy in those children deemed to have an unresectable tumor by diagnostic imaging.

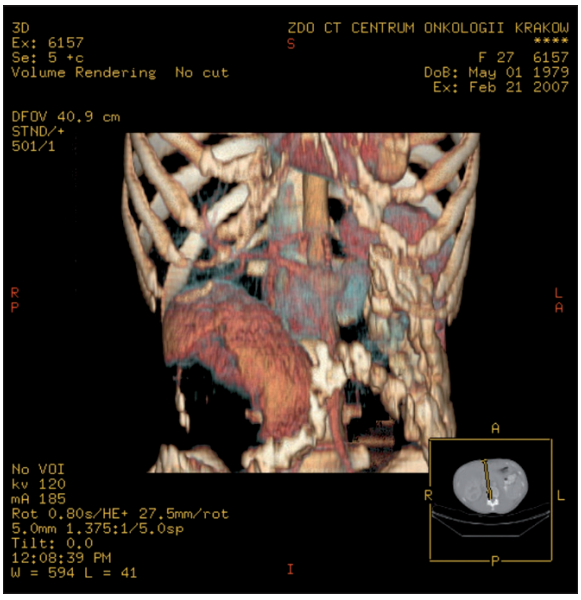

Figure 3. Abdominal CT – 3D reconstruction. Irregular tumor of the right kidney.

Therefore, we sougth to determine the degree of correlation between preoperative CT scan imaging of Wilms' tumor with the histopathological staging in our institution.

Case Report

A 28-year-old female patient presented to Oncology Institute with right kidney tumor. Her left kidney was resected due to Wilms' tumor in the sixth month of her life. Right kidney tumor was discovered, when she was 16 years old. In computed tomography examination the right kidney tumor of 70×35 mm in size was confirmed. The sample for histopathologic examination was taken and revealed epithelial type of nephroblastoma (standard form), medium grade malignancy. Due to the fact that partial nephrectomy was not considered to be an option, we tried to introduce radiation therapy toward kidney and the tumor up to the dose of 2400 cGy (the boost directed to the tumor 400 cGy/g). A partial tumor regression was obtain to the size of 4.4×3.2 cm. The patient was then qualified to right kidney partial resection. However, proposed treatment was not accepted by the patient.

After 12 years the patient come back complaining of pain in the lumbar part of the back. Abdominal ultrasound was performed and demonstrated a massive right kidney tumor. Prior to the surgical procedure, a CT of the abdominal was performed with the use of 16-slice CT scanner.

An imaging programme specific for renal arteries in layers 1.25 mm has been applied. 120 ml of nonionic contrast was administered. The speed of contrast flow was 4 ml/sek, using Smart prep programme for renal artery detection.

The examination revealed following findings:

,,Right kidney showing signs of compensative overgrowth, rotated and moved upwards, lies along with the visceral surface of the liver. In the lower and peripheral part of the

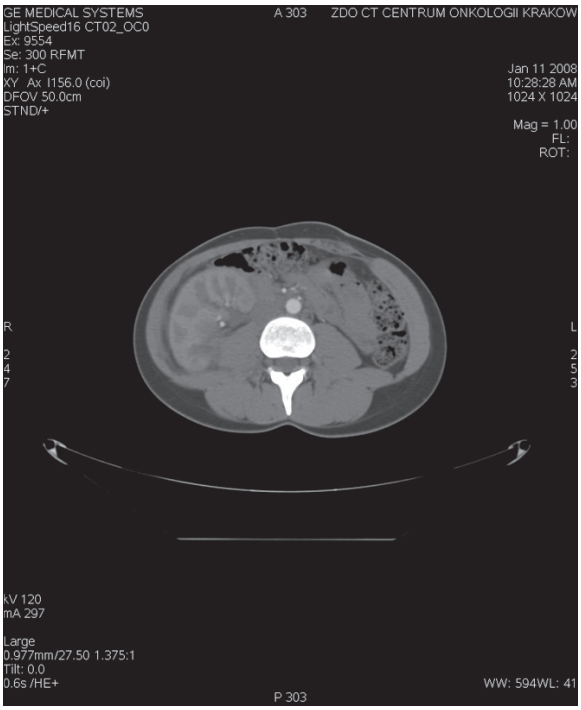

Figure 4. Post operative abdominal CT – post contrast phase. Axial image. Right kidney is preserved, with normal function. No recurrent tumor is visible.

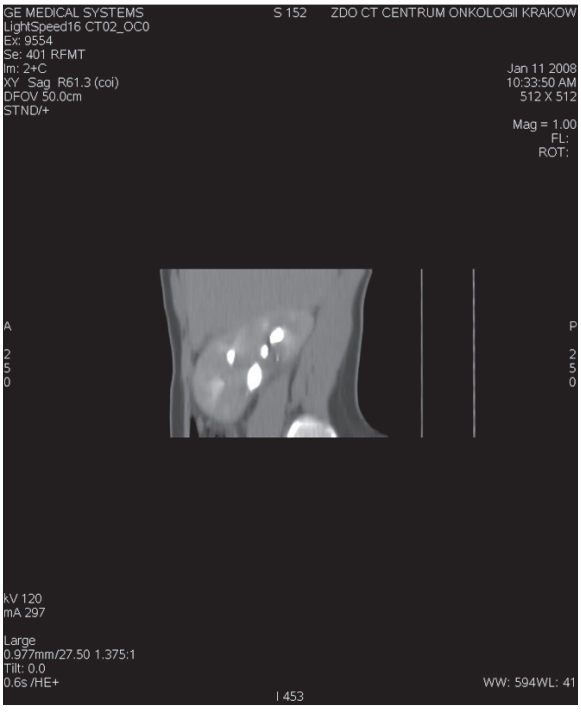

Figure 5. Post operative abdominal CT – post contrast phase. Saggital reconstruction. Right kidney is preserved, with normal function. No recurrent tumor is visible.

kidney a nodular mass 7×10×9 cm in size was visible. The tumor is well-demarkated, showing mixed attenuation, extends from the inferio-lateral kidney pole. The tumor is adjacent to psoas muscle and abdominal cavity walls; no infiltration of those structures is visible. Described lesion

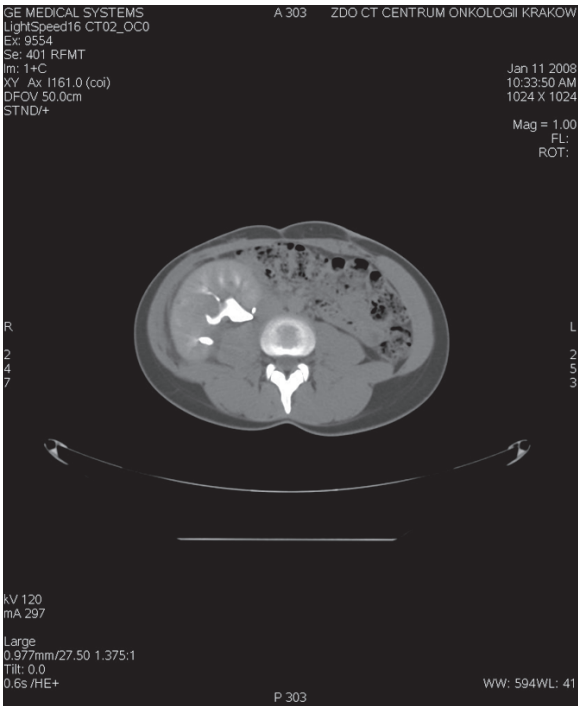

Figure 6. Post operative abdominal CT – post contrast late phase. Axial image. Right kidney is preserved, with normal function. No recurrent tumor is visible.

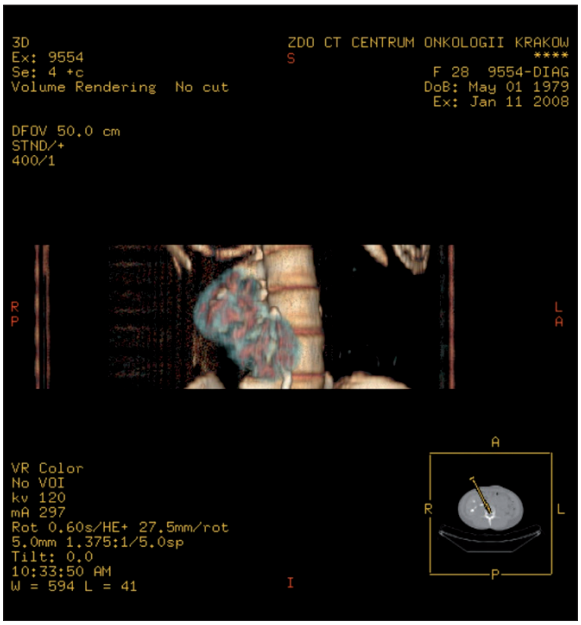

Figure 7. Post operative abdominal CT –3D reconstruction. Right kidney is preserved, with normal function. No recurrent tumor is visible.

surrounds inferior-medial part of the kidney and approaches kidney pelvis without infiltrating them. Superio-anterior and medial part of the kidney shows normal structure. Renal artery is preserved, extends posteriorly to the inferior vena cava. Tumor's vascularity seems to derive from segmental artery to the lower pole of the kidney. Collateral circulation is developed in the area surrounding the tumor. There are calcifications visible within tumor's capsule.

Lymph nodes enlargement within periaortal area was not detected (Figures 1–3).

Surgical procedure was performed resulting in excision of the tumor and saving the kidney. Histopathological examination revealed nephroblastoma (Wilms' tumor), with predominantly tubuler pattern. The tumor was surrounded by a thick, fibrous capsule. Surrounding parenchyma and fat tissue were not involved. Parenchyma beyond the tumor showed features of chronic inflammation.

CT examination performed 4 months after the surgical procedure revealed what follows: right kidney of 137×51 mm in size, normal location. Contours and thickness of the kidney parenchyma within normal limits. Cortex-medullary structure is preserved Calyces and pelvis of the kidney is not widened. The kidney produces urine normally (Figures 4–7).

Discussion

Wilms' tumor is the most common genitourinary tumor in childchood.

Bilateral Wilms' tumor (BWT) is rare, it accounts for 5% of all patients registered to the National Wilms' Tumor Study Group (NWSTG) [2].

Before the initiation of the large Wilms' tumor studies the NWTS group in the United States and SIOP in Europe, radical surgical ablation was thought to be essential for curing bilateral Wilms' Tumors. The approach is different now: NWTS is concentrated on primary surgery followed by adjuvant therapy consisting of chemotherapy with or whithout radiotherapy. SIOP is based on radiological staging with initial chemotherapy followed by surgery.

However, the ability of CT to determine the stage of a tumor accurately has been questioned by some authors [3]. We have tried to evaluate the accuracy of preoperative CT staging at our institution by correlation with histopathological findings.

On base of classical CT 2D axial it is difficult to estimate properly the local advance scan [4]. In our institution we examined the patient using 16-row computed tomograph. Than we generated 3D image of the renal tumor using GE Advantage Workstation. According to our protocol it was possible to estimate not only the size of the tumor, but also the infiltration of the renal capsule and lack of invasion to the renal pelvis. We also exclude of infiltration of vessels outside the kidney, regional extension into perirenal soft tissue and presence of the lymph nodes in renal hilus or periaortic chain. These findings enable surgery with preservation the part of the normal kidney. The tumor in our patient was correctly staged on the base of the CT (with 3D reconstruction with interval of 1.25 mm), which was proved in histological exam of the operated tumor.

Correct staging of the Wilms' tumor is crucial for choosing the proper therapy.

The strategies for optimal management have been studied extensively, and significant progress has been made in recent decades. K.W. Gow, I.F. Roberts [4] at all found, that it is difficult to differenciate between Stage I and Stage II,and a lot of tumors are overstaging to Stage II. Althought Stage I is intrarenal basing on CT examination, their data indicate that they are comparable in size with Stage II tumors. The results of overstaging this group is not clinically significant because both groups receive similar therapy.

The distinction between Stage II and Stage III is more important because the latter might be considered for primary chemotherapy if primary surgery is felt to be too hazardous. There are different protocols for chemotherapy between Stage II and Stage III Wilms tumors. Treatment of Stage III also includes radiotherapy. The consequences of over – or understaging are obvious, especially considering the potential side effects of radiotherapy such as intestinal stricture, ulceration, perforation, hematochezia or osteonecrosis [5,6].

When combined with adjuvant radiation and/or chemiotherapy, nephron sparing surgery provides an opportunity to preserve renal function for patients with bilateral Wilms' tumors [7].

Some authors emphasize that magnetic resonance imaging may be a usefull tool to determine diagnosis, capsular and vascular involvement [8]. However we think that CT exam is shorter (does not demand provides child sedation), and whole abdomen view (also bones) at the same time. Therefore, we would recommend looking for the best CT protocol for renal tumors staging.

Conclusions

Multislice abdominal CT visualized tumor of the single kidney very clearly, showed location of the tumor and no infiltration to the kidney pelvis or vessels, which allowed nephron sparing therapy.

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