Image of atypical Wilms tumor

Poster No.: C-1597
Congress: ECR 2019
Type: Educational Exhibit
Authors: M. D. C. Ojados Hernández¹, C. Fernández Hernández¹, I. Vicente Zapata¹, A. Gilabert Úbeda¹, C. Serrano Garcia¹, A. Navarro Baño²; ¹Murcia/ES, ²El Palmar/ES
Keywords: Abdomen, Paediatric, Kidney, CT, MR, Ultrasound, Diagnostic procedure, Education, Perception image, Cancer, Metastases, Education and training
DOI: 10.26044/ecr2019/C-1597

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR's endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

- Describe the key findings of the classic Wilms tumor.
- Analyze the atypical Wilms tumor’s presentation.
- Describe the key findings of the atypical Wilms tumor’s presentations in ultrasound, CT and MR.
Background

Wilms tumor, or nephroblastoma, is the most common renal solid mass and abdominal malignancy of childhood and occurs in approximately 1: 10,000 persons. Its peak incidence is at 3-4 years of age, and 80% of patients present before 5 years of age. Is uncommon in neonates, less than 0.16% of the cases.

Wilms tumor is most frequently sporadically, up to 90% of cases. On other occasions, it occurs in hereditary forms associated with other congenital anomalies such as cryptorchidism, hemihypertrophy, hypospadias, and sporadic aniridia. The genetic syndromes that present Wilms tumor are the syndrome WARG, Drash syndrome, and Beckwith-Wiedemann. Chromosome 11 is involved in these less frequent forms. Family Wilms tumor is rare, 1%, and is not associated with mutations on chromosome 11.

The common histology of the tumor is a triphasic appearance consisting of stromal, epithelial and blastemal elements that recapitulate the normal development of the kidney. But, in some cases, they may have a monophasic or biphasic appearance. Less frequently, unfavorable histologies are found such as teratoid and anaplastic forms. The teratoid form has different tissue lines to the kidneys, such as muscle, bone or cartilage. In 10% of the cases, Wilms tumor shows anaplastic histology. Anaplastic and teratoid histology is considered unfavorable with a poor prognosis.

The typical presentation of the Wilms Tumor is asymptomatic abdominal mass. In other cases, it can be presented as anemia, hypertension, fever or hematuria.

The presentation form in the debut of most Wilms tumor are solitary lesions, but at least 12% of patients can develop a multifocal tumor in the same kidney. Approximately 6% of patients present synchronous bilateral renal tumor.

Screening for suspected abdominal tumor is performed with US, but US is operator-dependent with a limited field of view in addition to other technical limitations. The evaluation with magnetic resonance (MR) imaging or computed tomography (CT) is generally necessary for optimal staging and operative planning.

The appearance of Wilms tumor at US is variable. Usually, the tumor is heterogeneous with hypoechoic and anechoic areas representing hemorrhage, necrosis, and cysts. Calcifications are seen in 9% of patients. US with color Doppler imaging is a sensitive modality for the tumor spread into the inferior vena cava (IVC).
At CT, the tumor usually appears as a large, heterogeneous, intrarenal mass that enhances the normal renal parenchyma adjacent. Hypoattenuating foci of necrosis and old hemorrhage are common, and fat attenuation may also be seen. Calcifications are seen in approximately 15% of cases.

At MR imaging, the tumor is typically heterogeneous, lobulated, and hypointense compared with the kidney on T1-weighted images and hyperintense on T2-weighted images. Foci of increased signal intensity on T1-weighted images representing hemorrhage may be observed. After administration of gadolinium, the tumor enhances less avidly than the adjacent kidney.

Preoperative diagnosis and radiological follow-up are crucial for the success of the therapy. Surgical resection is mandatory for staging and treatment. Staging of Wilms tumor is eminently surgical. The National Wilms Tumor Studies (NWTS) classifies the tumor in V stages.

Although Wilms tumor is a malignant tumor, the prognosis is excellent with multiple therapy (surgery, chemotherapy and radiotherapy). The 5-year survival rate of 90%, reaching 95% in tumors <4 cm, and 70% for metastatic disease.
Findings and procedure details

Wilms tumor is usually a solitary lesion in a kidney and there is usually no metastasis at the time of diagnosis, although throughout the disease can develop lung metastases, liver and rarely in bone and brain. Occasionally the tumor extends intravascularly to the renal vein and inferior vena cava (ICV).

Below we present the less frequent presentations of Wilms tumor, showing the most representative images in each case.

**Bilateral Wilms tumor.**

Usually, Wilms tumor is a single lesion in a single kidney. They are large lesions due to the insidious growth of the lesion and can cross the midline. However, up to 6-10% of cases can be bilateral.

The contralateral kidney needs to be carefully screened. The presence of a synchronous lesion changes the stage to stage V, and this will change the therapeutic approach.

The staging of the bilateral tumor must be done independently for each kidney, and the prognosis depends on the higher individual stage.

The unilateral Wilms tumor is usually treated with nephrectomy followed by adjuvant chemotherapy. In bilateral tumor, preoperative chemotherapy is important, since the current approach is resection of the tumor with preservation of the normal parenchyma.

**Intravascular thrombus**

It has been reported that intravascular thrombus occurs in 20 to 35% of Wilms Tumor, while IVC involvement is reported in only 4 to 10%. A thrombus that reaches the atrium occurs in less than 1% of all patients.

Children with WT and a thrombus in their IVC are usually asymptomatic. This is probably due to the persistence of venous blood flow despite the presence of a tumor thrombus within the lumen of the cava.

When the thrombus is below the hepatic veins, management options include initial surgery or neoadjuvant chemotherapy followed by delayed surgery. With greater extension of the proximal thrombus and in stable patients, neo-adjuvant chemotherapy has gained consensus as the primary approach of choice regardless of the protocol adopted. In those cases in which there is thrombus in the IVC and right atrium, in
which surgery is decided as the first option, cardiopulmonary bypass for removal is required. Adherent ICV thrombus has been treated by different surgical procedures. Anticoagulation is not indicated routinely.

The presence of thrombus tumor represents a major surgical risk. The preoperative identification, the determination of extension and the possible invasion of the IVC wall are crucial factors in the planning of the therapy. It must be identified for decreases the rate of surgery complications.

Therefore, the presence of thrombus in the ICV is not a prognostic factor, not decrease survival rate, however the patient's morbidity increases due to the risk of pulmonary thrombosis.

It is crucial to identify thrombus in imaging studies because changes the therapeutic plan. Not identifying the intravascular thrombus in pre-surgical studies increases complications during surgery.

The technique of choice for the detection of thrombus is the US with color Doppler because it sometimes goes unnoticed in CT studies. MR is also an ideal technique for the identification of the thrombus and its extension. The performance of presurgical echocardiography is important to detect the presence of thrombus in the right atrium.

**Metastatic lung disease**

The lung is the most common site of metastatic disease in Wilms Tumor (85% of cases of metastatic disease). 10% of patients have pulmonary metastases at diagnosis. The 5-year survival rate decreases to 20% -70%.

The traditional diagnostic method to detect metastatic disease in the lung in children with Wilms tumor is chest radiography. The use of chest radiography versus CT for evaluation for lung metastasis at diagnosis remains controversial. The results obtained from the most of studies are based on the use of radiography. However, CT is more sensitive for small nodules. There are studies that shown no substantial differences in the outcome after the administration of local therapy of those who were treated as a metastatic disease in those patients with pulmonary metastases only visible by CT. However, CT is essential for those with unfavorable histology or stage III.

**Relapse**

Approximately 15% of primary tumors with favorable histology and 50% of cases of high-risk tumors develop relapse. Approximately 95% of tumor recurrences occur within 2
years of the initial diagnosis. The long-delayed recurrence, > 5 years after diagnosis, is very infrequent, only 0.5% of cases.

The most frequent relapse sites are the lung, abdomen and less frequently bone and brain.

Recurrent tumors are defined as very high risk to develop a chemoresistant disease, which correlates with a poor prognosis.

In response to a series of items, patients with Tumor Wilms were classified into 3 risk categories for recurrence:

- **Standard risk (30% of cases of recurrence):** patients with favorable-histology WT with relapse after therapy with only vincristine and/or actinomycin D. Free survival range 70-80% range.
- **High-risk (45-50% relapse):** with favorable-histology WT with relapse after therapy with three or more agents. Free survival range 40-50%.
- **Very-high-risk (10-15% relapse):** patients with recurrent anaplastic or blastemal-predominant WT. Free survival range 10%.

**Pulmonary relapse**

The most Wilms tumors recurrences occur in the lung, 58% of cases.

The sites of pulmonary relapse included in the lung parenchyma, pleura, and less frequent mediastinum. The 3-year survival rate decreases to 45% in those patients with Wilms tumor with relapse confined to the lung. The chest X-ray is, until now, the test of choice for the detection of lung disease. CT helps identify a subset of patients with local disease who have an increased risk of relapse of the lung.

**Abdominal relapse**

Abdominal relapse is uncommon. The incidence of local recurrence is widely reported in the literature and it varies from 7-20%. This rate drops to about 3% in small tumors.

While Tumor Wilms is generally favorable in patients with abdominal relapse carries a worse prognosis. Decreasing the 3-year survival rate to 28-30%.

The most frequent sites of abdominal relapse included the kidney, peritoneum and lymph nodes. Relapse in the surgical bed is uncommon, being the less frequent site of relapse in the published articles.
The patients with highest risk of abdominal relapse are those with unfavorable histology, nephroblastomatosis, spillage tumor, the inflammatory pseudocapsule and tumors > 4cm.

It's recommended to closely follow-up with imaging studies of high-risk patients. Ultrasound is sufficient for routine monitoring. It is suggested that high-risk patients be examined every 3 months during the first 3 years. The CT and MR should be used for uncertain findings.

**Multifocal disease**

It's estimated that the risk of multifocal Wilms tumor is as high as 15%, but it is much lower among patients with smaller and lower stage tumors.

Patients with nephroblastomatosis have a high risk for the development of multifocal cancer. These patients should be monitored closely during the first 3-years after diagnosis due to high risk of recurrence.
Fig. 1: Abdominal radiography in Wilms tumor.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
**Fig. 2:** Ultrasound images in bilateral Wilms tumor.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
**Fig. 3:** Ultrasound images in bilateral Wilms tumor.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia

Transverse ultrasound image (A) and transverse color Doppler image (B). Bilateral Wilms tumor. Large, well-defined, heterogenous renal mass (blue asterisk) with hypoechogetic with cyst areas representing hemorrhage or necrosis. Presence of color Doppler flow in the tumor mass (B).
Fig. 4: Bilateral Wilms tumor and thrombus in IVC.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
Axial fat-sat T2-weighted MR image (A), axial enhanced fat-sat T2-weighted MR image (B) and coronal fat-sat T2-weighted MR images (C, D). Bilateral Wilms tumor. Heterogeneous bilateral mass (blue asterisk) isointense, compared with the adjacent kidney, with a foci of decrease signal representing necrosis or hemorrhage. Tumor enhances less avidly than adjacent kidney. Post-surgical image after bilateral partial nephrectomy (D).

**Fig. 5:** Bilateral Wilms tumor.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
**Fig. 6:** Wilms tumor IV stage, pleural metastasis.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
Fig. 7: Wilms tumor. Pulmonary relapse.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
Coronal enhanced fat-sat T2-weighted MR image (A, C) and coronal T2-weighted MR image (B). Left Wilms tumor with abdominal relapse in nephrectomy bed. Large tumor mass in left kidney (blue asterisk) with heterogeneous appearance and necrosis or hemorrhage foci. Post-surgical image after nephrectomy (B). Solid mass with homogenous enhanced and necrotic center at the site of the surgical bed (green asterisk).

**Fig. 8:** Wilms tumor. Abdominal relapse.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
Fig. 9: Wilms tumor IV stage with hepatic and pulmonary metastasis.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
**Fig. 10:** Wilms Tumor. Pulmonary relapse.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
**Fig. 11:** Wilms tumor IV stage. Debut with hepatic metastasis rupture.

© Departamento de Radiología. Hospital Universitario Virgen de la Arrixaca. Murcia
Conclusion

Wilms tumor is the most common renal solid mass and abdominal malignancy of childhood. Although Wilms tumor is a malignant tumor, the prognosis is excellent with a 5-year survival rate of 90%, and 70% for metastatic disease.

The classic Wilms tumor’s presentation is usually a solitary lesion in a kidney and there is usually no metastasis at the time of diagnosis. Although the disease can develop lung metastases, liver and rarely in bone and brain. However, there are other atypical forms of Wilms tumor presentation as a bilateral tumor, multifocal tumor, metastatic disease at diagnosis, intravascular tumor, pulmonary and abdominal relapse in which the survival rate and the success of the therapy can be compromised.
• Patterns of abdominal relapse and role of sonography in Wilms tumor. Pediatric Hematology and Oncology, 19:2, 107-115. DOI: 10.1080/08880010252825696