

## Casos Clínicos / Radiological Cases Report

### Bilateral Extrarenal Wilms' Tumor: Case Report

Teresa Proença e Cunha<sup>1\*</sup>, Isabel Duarte<sup>1</sup>, José Venâncio<sup>1</sup>, Nuno Jalles Tavares<sup>3</sup>, Saudade André<sup>2</sup>, Maria de Lurdes Orvalho<sup>1</sup>

Departments of Radiology<sup>1</sup> and Pathology<sup>2</sup>, Portuguese Institute of Oncology, Lisbon  
Nuclear Magnetic Resonance Unit<sup>3</sup>, Caselas, Lisbon

#### Resumo

Os autores descrevem um caso clínico de tumor de Wilms' extrarrenal de localização ao retroperitoneu, num doente do sexo feminino de 13 anos de idade. A revisão da literatura efectuada revelou a extrema raridade da apresentação bilateral extrarrenal numa criança mais velha.

#### Palavras-chave

Tumor de Wilms' Extrarrenal; Tumor de Wilms' Bilateral; Nefroblastoma.

#### Abstract

A case of an extrarenal Wilms' tumor arising in the retroperitoneum of a 13-year-old-girl is described. The review of the literature performed showed the extreme rarity of such a bilateral presentation in an older child.

#### Key-words

Extrarenal Wilms' Tumor; Bilateral Wilms' Tumor; Nephroblastoma.

The extrarenal location of Wilms' tumor is extremely rare [5,7], and even rarer when the tumor is bilateral. During childhood, the extrarenal Wilms' tumor usually presents itself as a solid, well defined and heterogeneous retroperitoneal mass[7], with necrosis and hemorrhagic areas. Nowadays, its histogenesis and embriogenesis are being discussed and the prognosis is not well known due to the small number of reported cases[7].

The main interest of this report lies in the initial diagnosis in an older child and on the bilaterality of the tumor at presentation.

#### Case Report

A 13-year-old girl had a one-day history of crampy abdominal pain with vomiting, constipation and temperature of 37.5°C. On physical examination a large abdominal swelling mass was noted over the superior and left abdominal quadrants, smooth, nontender, non-pulsatile and fixed. The remainder of the physical

examination revealed no other abnormal findings. The blood pressure and the laboratory investigations were normal. Abdominal ultrasound showed two large solid and well-circumscribed masses, located laterally to the spine (Fig. 1) and retroperitoneal blood vessels. These tumors extended from the superior abdomen down to the iliac crest and appeared heterogeneous, with several hypoechoic areas. The right mass displaced the inferior vena cava (IVC) anteriorly, the right kidney downward and the liver medially. On the left side, the kidney was posteriorly displaced (Fig. 2). No solid viscera was invaded by the tumors. On CT these aspects were confirmed (Fig. 3). There was no evidence of abdominal lymph nodes or liver metastases. MRI was performed to better localize the tumors and to further characterize its components. Several hypersignal areas on both T1- and T2-weighted images were consistent with hemorrhagic component, and areas of low-signal intensity on T1-weighted images and high-signal intensity on T2-weighted images evoking necrosis were also demonstrated. The solid part of the tumors appeared of intermediate signal intensity on both sequences. MRI corroborated solid but partially necrotic and hemorrhagic lesions. Coronal images showed the relationship of the aorta and IVC to the

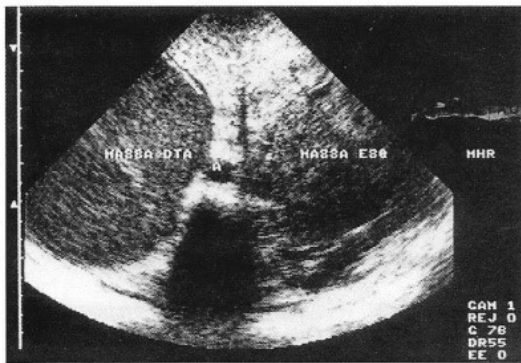


Fig. 1 - Supine transverse ultrasound scan shows two large solid masses laterally to the spine.



Fig. 2 - On a sagittal ultrasound scan, the left kidney was posteriorly displaced by a large solid mass. Note the well - demarcated interface between the kidney and mass.

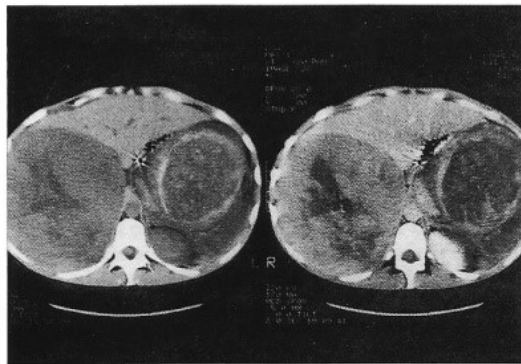


Fig. 3 - CT scan of the upper abdomen shows two large masses with a mixed pattern and heterogeneous enhancement with intravenous iodinated contrast material. They are separated from the liver and left kidney and adrenal gland.

tumors as a displacement without invasion (Fig. 4) and T2-weighted images documented a peripheral hyposignal ring separating the masses from the adjacent organs (Fig. 5).

Two biopsies of the masses were performed before surgery to differentiate retroperitoneal sarcoma from



Fig. 4 - Coronal T1-weighted image shows the relationship of the tumors to the IVC and the aorta. The proximal end of the left renal artery and the distal end of the left renal vein are patent.

lymphomatous masses of non-Hodgkin's lymphoma which indicate medical treatment. The biopsies revealed an undifferentiated malignant neoplasm. However, cytologic samples pointed sarcomatous components.

Two months later, at surgery two retroperitoneal tumors were found. The left mass was adherent to the stomach superiorly, to the colon medially and posteriorly to the kidney and pancreatic tail. The right mass invaded the upper pole of the kidney and the capsule of the inferior surface of the right liver lobe. The masses were removed with the spleen, left adrenal gland, left kidney, upper pole of the right kidney, pancreatic tail as well as parts of the colon, liver and stomach. During surgery it was necessary to perform a thoracotomy and to remove a portion of the left part of diaphragm. The macroscopic appearance of the right tumor was an ovoid rubbery mass, capsulated, measuring 13 cm at its widest point, with scattered zones of hemorrhage and necrosis. On cut section, yellow and firm areas were also identified. The left mass infiltrated the left kidney capsule; it was extensively necrotic and the larger section was about 14 cm. The spleen, colon, pancreas and left adrenal gland were not invaded by the tumor. The two neoplasms arose almost entirely in perirenal soft tissues without neoplastic infiltration of the kidneys. The left part of the resected diaphragm was involved by tumor. Microscopically the tumors were composed of residual nodules of renal blastema, tubules

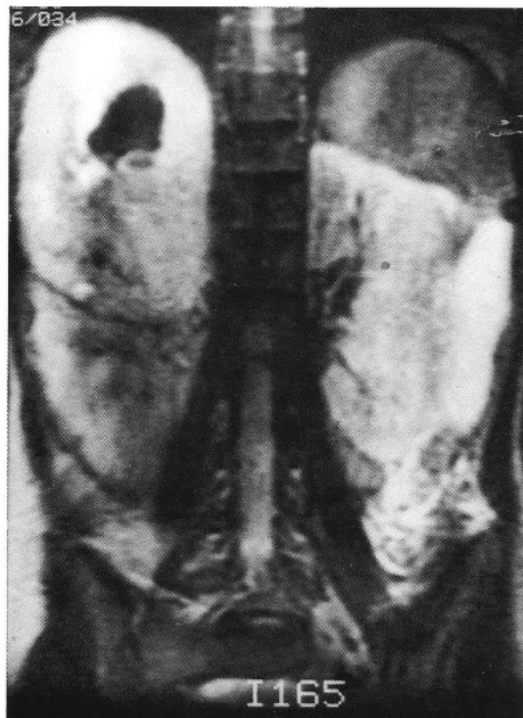


Fig. 5 - T2-weighted coronal MR image shows a peripheral hypo-signal ring separating the right mass from the adjacent organs. The central area of high-signal indicates necrosis and the hypo-signal area in its interior represents an extensive fibrous component.

and glomeruli, surrounded by bland fibrous and necrotic tissue. Final histologic diagnosis was extrarenal Wilms' tumors. Examination of the submitted lymph nodes showed no evidence of metastatic spread.

Preoperatively the patient received intravenous chemotherapy with Cis-Platinum, Vincristine, Doxorubicin, Actinomycin-D and Ifosfamide, with poor response on tumor size but with an increase of the necrotic component. Postoperatively, as a consolidation treatment, the patient was placed on external radiation therapy to the left kidney bed (30 Gy) as well as chemotherapy with Actinomycin-D, Vincristine, Epi-Doxorubicin and Ifosfamide.

On follow-up study, the girl is developing normally nine months after surgery. There is no evidence of abdominal mass, and chest roentgenogram shows no abnormalities. General physical examination is within normal limits.

## Discussion

Extrarenal Wilms' tumor is extremely rare and occurs more commonly in children[5,10]. In 1987, there were 17 reported cases and 11 occurred in children under 15[5] and in 1993 there were 34 cases described[8]. The retroperitoneum adjacent to the kidney and inguinal canal are the usual sites of origin[5,9], and

histologically they are indistinguishable from Wilms' tumors in the kidney[10].

A number of authors have reported nephroblastic tissue in teratomas, usually in retroperitoneal or sacrococcygeal locations[5] and this has led to discussion on the embryologic origin of this kind of tumors. Most authors believe that true extrarenal Wilms' tumors arise from a persistent fetal metanephric blastema[7] that failed to differentiate into normal tubules and glomeruli[3], but others argue that nephroblastoma is merely a component of a teratoma, and thus derived from a primitive mesodermal or "totipotent" cell [5,6]. Therefore complete examination of these "tumors" is mandatory to exclude a primary teratoma with predominance of nephroblastic elements[1,5,7].

According to Koretz, there are two important issues in extrarenal Wilms' tumors: 1) the possibility of having missed a supernumerary kidney as the site of origin[1] and 2) incomplete tissue examination that failed to detect extrarenal teratoma tissue[5]. In this case normal kidneys were found on the histologic examination and no nephroblastic tissue was found on complete microscopic examination of the surgical specimen. The histologic consequence of preoperative chemotherapy was noted to be necrosis predominantly of the undifferentiated tissue.

The differential diagnosis includes retroperitoneal sarcoma and neuroblastoma. Definitive diagnosis of the tumor is supported by surgical and histological findings.

Extrarenal Wilms' tumor has the potential for local recurrence and distant metastasis, but with current therapy, good results can be expected even in patients with incomplete surgical excision of their tumors[4]. The imagiologic evaluation, with different techniques (US, CT, MRI) shows the inhomogeneous character of the tumors[2]. They can correctly identify the hemorrhagic, necrotic and solid parts of the tumors and the pseudocapsule around the masses was particularly well visible on MRI and is probably composed of compressed renal parenchyma[2]. There was no specific imagiologic feature that could differentiate Wilms' tumor from other retroperitoneal neoplasms, such as sarcoma and neuroblastoma.

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## Correspondence

Teresa Proença e Cunha  
Departamento de Imagiologia  
Instituto Português de Oncologia - Lisboa  
Rua Prof. Lima Basto  
1093 Lisboa Codex