

## Giant relapsed Wilms tumour with liver and lung metastases in a child: a case report

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**Section:** Paediatric radiology

**Area of Interest:** Abdomen

**Procedure:** Diagnostic procedure

**Imaging Technique:** CT

**Imaging Technique:** Ultrasound

**Imaging Technique:** Ultrasound-Colour Doppler

**Imaging Technique:** Conventional radiography

**Special Focus:** Metastases Neoplasia Case Type: Clinical Cases

**Authors:** Eliza Stavride, Katerina Manavi, Marianna Theodorou, Melpomeni Kosmidou, Antonios Theodorakopoulos, Ioannis Tsitouridis

**Patient:** 6 years, male

### Clinical History:

A 6-year-old boy was referred from another hospital with a 2-day history of abdominal pain, fever, pallor and weakness. Examination revealed a large solid left upper quadrant mass. The history was unclear. Blood tests revealed low blood haemoglobin (4.9 mg/dl). He was transfused and transferred to our hospital for further work-up.

### Imaging Findings:

Chest X-ray demonstrated elevation of the left diaphragm and absence of the gastric bubble. At this stage, further history of left nephrectomy and postoperative chemotherapy due to nephroblastoma at age 2 was elicited, with further relapse a year later when he underwent excision of a mass and subsequent irradiation and chemotherapy. He had had no follow-up for the last 16 months. CT revealed a huge, well-defined, heterogeneously enhancing mass, measuring 15x10x13cm, and probably arising from the left renal bed. Central hypodense areas likely represent necrosis and hemorrhage. Multiple metastatic liver lesions and 3-4 metastatic lung nodules were also demonstrated. Ultrasound, performed to investigate the patency of the vessels, demonstrated a heterogeneously echogenic mass with hypoechoic lesions and mild intratumoral vascularity. The mass was compressing the spleen and the stomach.

### Discussion:

Wilms tumour (WT), also known as nephroblastoma, is the most common malignant renal tumour in childhood and the second most common malignancy of the abdomen after neuroblastoma [1, 2, 3]. Most children are diagnosed between the ages of 1 and 5 years [1, 2]. It usually originates from the renal parenchyma, but it can extend beyond

the renal capsule. In rare cases it can arise from extra-renal sites such as the retroperitoneum, inguinal canal, scrotum and vagina, and is called extra-renal [4, 5, 6]. About 10-13% of WT are associated with additional abnormalities and exist as a part of a congenital syndrome [2, 7]. WT can spread to abdominal lymph nodes and haematogeneously metastasise to the lungs and liver [2, 4]. It can also invade blood vessels and extend into the renal vein [2].

WT classically appears with a painless abdominal swelling in a healthy looking patient [5]. Other clinical symptoms include hypertension, abdominal pain, haematuria and fever [8]. US is usually the first imaging modality used and in most of the cases it demonstrates a large, solid, well-marginated, heterogeneous mass with internal hypoechoic lesions representing haemorrhage, necrosis or cystic degeneration. It can also contain calcification and fat. CT is then used to evaluate the extent of the tumour and detect metastatic lesions to nodes, liver and lung. CT shows a large, solid, well-defined mass of heterogeneous density and sometimes central hypodense areas of necrosis or haemorrhage. MR imaging is rarely used to detect tumour recurrence [8]. Diagnosis must be confirmed histologically by biopsy or surgical specimen. Three tumour components are usually seen: blastema, epithelia and stroma [3].

Over 85% of WT cases are treated by a combination of surgery, radiotherapy and chemotherapy, depending on the histologic cell type of the tumour [9]. The presence of an anaplastic component is associated with the development of metastases and recurrence after surgical excision [8, 9]. Relapses occur in 15-20% of patients and most of those are within two years of diagnosis. WT can recur locally or in several other sites, most commonly the abdomen, lungs, and contralateral kidney. The latter is associated with a favourable outcome [9, 10]. Regular surveillance with US or CT for three years after completion of treatment is used to detect most abdominal recurrences [11].

**Differential Diagnosis List:** Relapsed nephroblastoma (Wilm's tumour) with multiple liver and lung metastases, Neuroblastoma, Multilocular cystic renal tumour, Clear cell sarcoma, Renal rhabdoid tumour, Renal abscess, Angiomyolipoma, Renal cell cancer

**Final Diagnosis:** Relapsed nephroblastoma (Wilm's tumour) with multiple liver and lung metastases

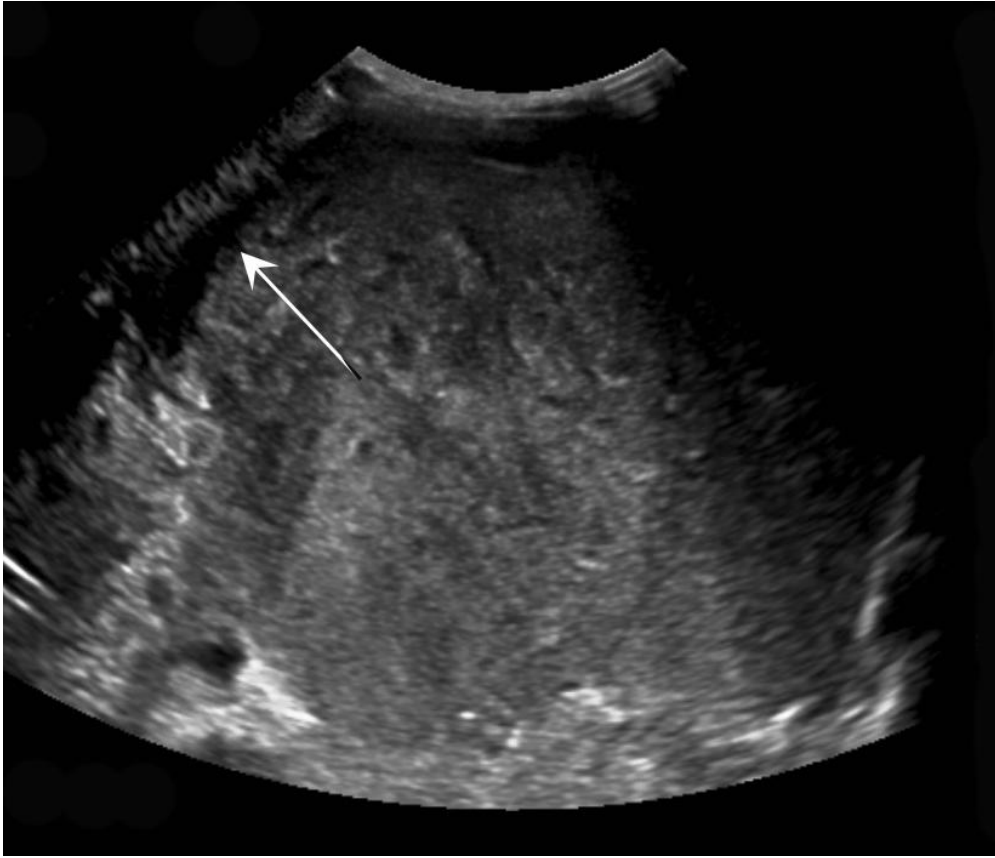
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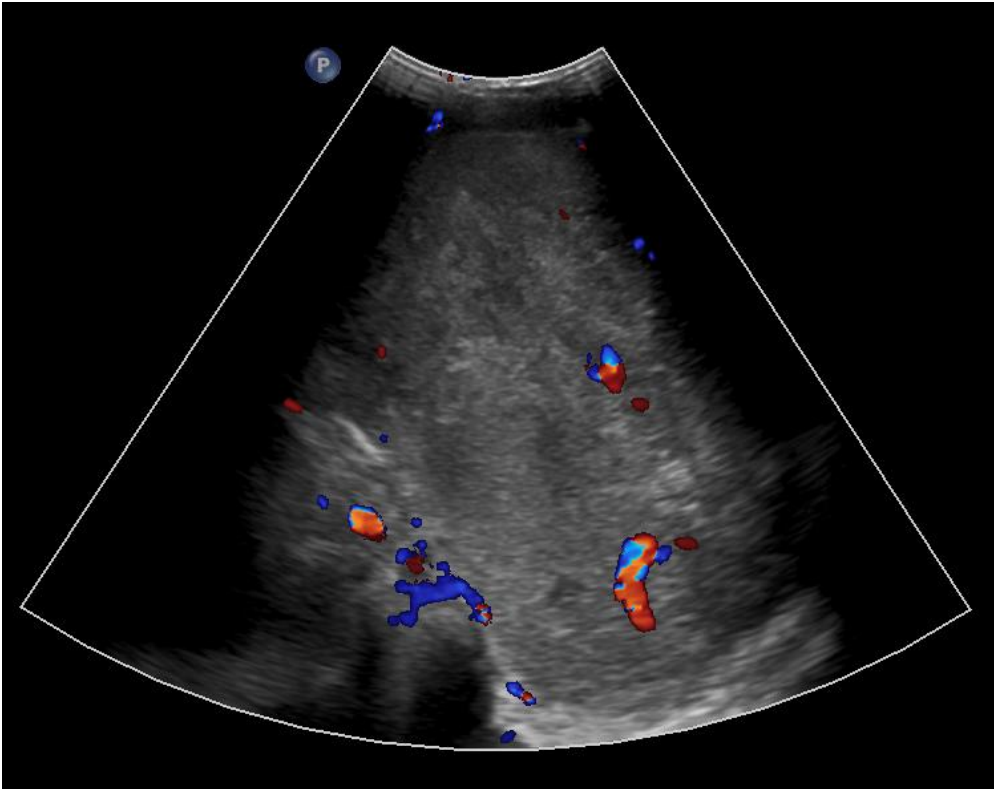
**Figure 1**

a



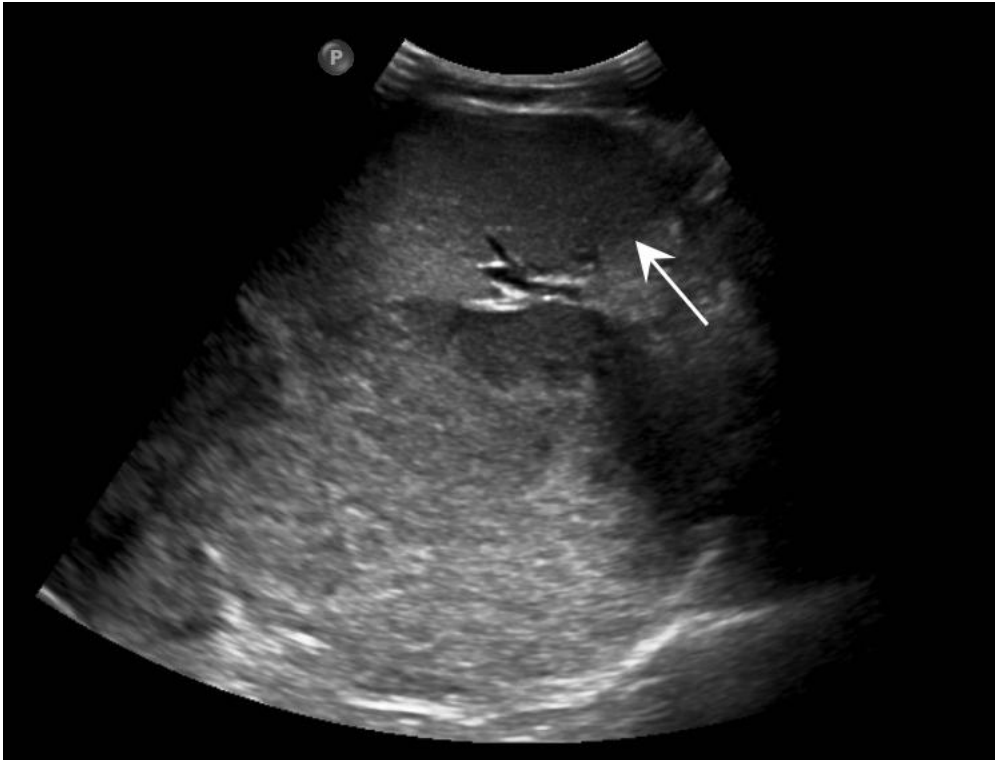
**Description:** Ultrasound imaging showing a large heterogeneously echogenic mass with hypoechoic areas representing necrosis or haemorrhage. The mass is compressing the stomach (arrow). **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

**b**



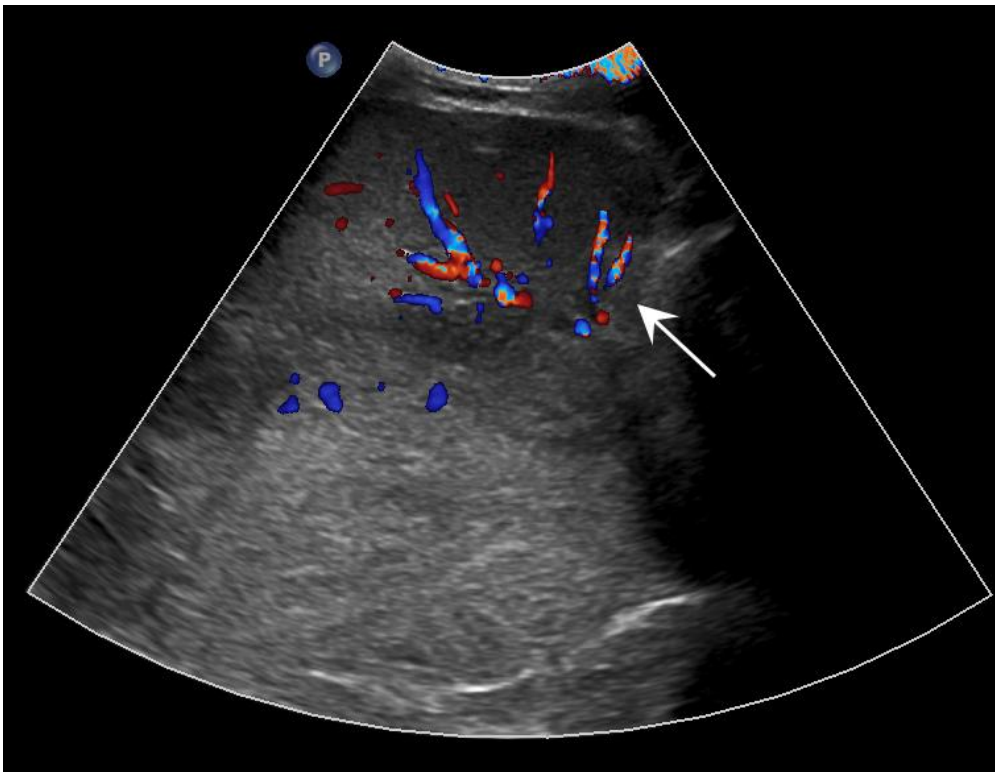
**Description:** Colour Doppler ultrasound imaging showing a large heterogeneously echogenic mass with mild intratumoral vascularity. **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

c



**Description:** Ultrasound imaging showing the mass compressing the spleen (arrow). **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

d



**Description:** Colour Doppler ultrasound imaging showing the mass compressing the spleen. **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

**Figure 2**

a



**Description:** Axial unenhanced computed tomography image showing a huge well-defined mass located between the liver and the spleen. **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

**b**



**Description:** Axial contrast-enhanced computed tomography image showing mild enhancement and hypoattenuating areas of necrosis or haemorrhage. **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

**c**



**Description:** Axial contrast-enhanced computed tomography image showing the heterogeneous mass compressing the stomach (arrow). **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece



**Figure 3**

a



**Description:** Sagittal contrast-enhanced image showing a huge mass arising from the retroperitoneum and occupying almost the whole abdomen. **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

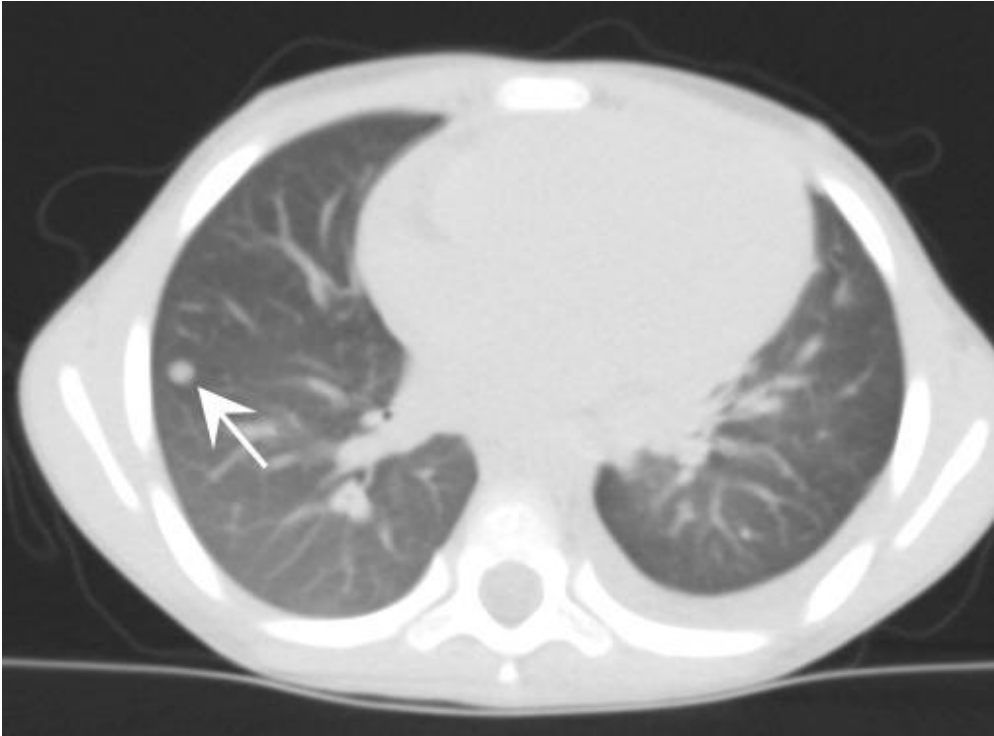
**b**



**Description:** Coronal contrast-enhanced image showing a large mass (arrows) with hypodense internal areas lying between the liver and the spleen and elevating the left hemidiaphragm. **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

**Figure 4**

**a**



**Description:** Chest computed tomography image showing the most typical metastatic nodule in the right lung (arrow). **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

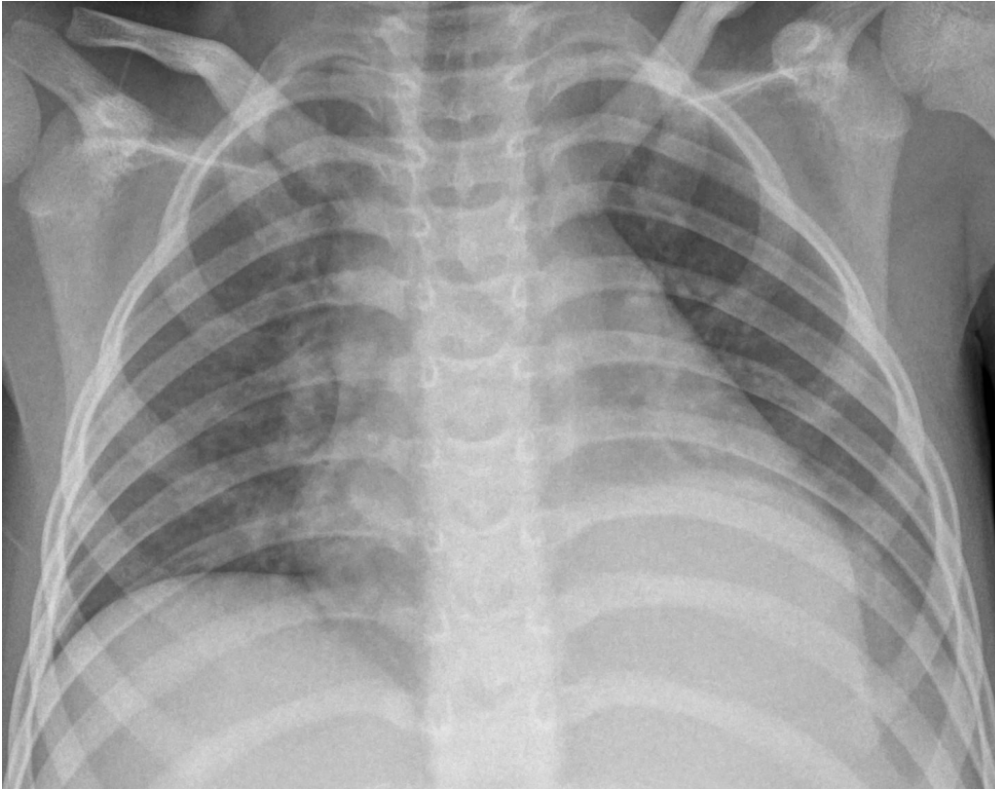
**b**



**Description:** Axial contrast-enhanced computed tomography image showing at least three hypoattenuating round lesions, representing liver metastases (arrows). **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

**Figure 5**

**a**



**Description:** A chest radiography demonstrating elevation of the left hemidiaphragm and absence of the gastric bubble. **Origin:** Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece