

Case report

DOI: <http://doi.org/10.4038/slju.v13i0.4070>

A case of adult Wilm's tumour masquerading as an inflammatory mass in the kidney

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Case report

A 20-year old girl having right sided abdominal pain became faintish and was admitted to the local hospital. Abdominal ultrasonography suggested a renal mass and associated hydronephrosis possibly secondary to sepsis but the possibility of a neoplasm could not be excluded. There was no history of weight loss or haematuria. Physical examination showed a large, firm, ballotable and tender mass on the right upper quadrant of abdomen of an ill-looking girl. Computed tomography showed a well-defined heterogeneous lesion with contrast enhancement in the right hydronephrotic kidney (Figure 1 and 2). The mass had a predominantly solid component and scanty cystic areas and was 10 cm × 9 cm in size. There was no renal vein invasion or lymphadenopathy. Ultrasound guided core needle biopsy was done to ascertain the nature of the mass and the histology showed a malignant tumour suggestive of a nephroblastoma.

Right sided radical nephrectomy was performed transperitoneally and the histology confirmed the tumour to be a nephroblastoma (Wilms' tumour) with favourable histology. The tumour composed mainly of epithelial component, without anaplastic elements.

Seven lymph nodes removed were negative for tumour cells. The tumour was invading the perirenal fat and hence it was classified as stage II (NWTS staging). She completed a course (three weekly, 12 cycles) of chemotherapy comprising of etoposide, carboplatin, cyclophosphamide and doxorubicin. She also had 15 days of external beam radiotherapy after the first cycle of chemotherapy. She is asymptomatic and free of any recurrences five years after surgery.

Discussion

Nephroblastoma, also known as Wilms' tumour is a malignant embryonal neoplasm derived from nephrogenic blastemal cells that both replicates the histology of developing kidneys and often shows divergent patterns of differentiation. 98% of Wilms' tumours occur in children under the age of 10 years (1). Commonest presentation is an incidentally found

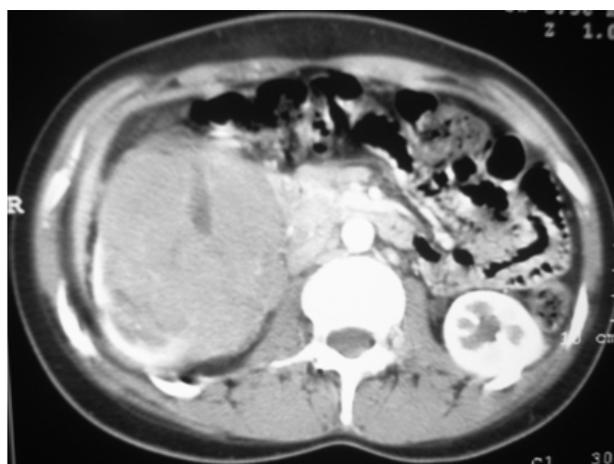


Figure 1.



Figure 2.



asymptomatic abdominal mass by the parents of the child. However acute abdomen like features due to tumour rupture may be the presentation

Wilms' tumour is the most common malignant tumour of the urinary tract in children but is rare in adults (over the age of 15 years). The preoperative diagnosis of adult Wilms' tumour is extremely difficult because there are no radiographic investigations, especially characteristic CT features that can distinguish it from the more common renal cell carcinoma. It is interesting to note that another patient with adult Wilms' tumour has had hydronephrosis in addition to the tumour mass in the CT scan, a finding which was seen in our patient as well (2). Whether this could be a useful feature to differentiate an adult Wilms' tumour from other renal neoplasia preoperatively needs more evidence. Commonly found renal cell carcinoma does not produce hydronephrosis even when big.

Percutaneous needle biopsy generally is used to assess the nature of a doubtful renal mass or a small renal mass where characteristic radiological features of a malignant renal neoplasm are not apparent. It has been shown to be effective in approximately 90% of cases in the hands of experienced radiologists (3). As there was a doubt as to the possibility of an inflammatory lesion of the kidney or a rare neuroendocrine tumour the ultrasound guided biopsy was useful in the preoperative diagnosis of a nephroblastoma in this adult patient.

The true incidence of adult Wilms' tumour is uncertain because of variations in terminology and pathologic criteria. Whether there are differences between the tumour in adults and children is unclear. Microscopically, there is no difference. Wilms' tumour, whether it occurs in childhood or in adult life, does not contain the mature glandular elements of renal carcinoma, but the findings of abortive glomerular or tubular (epithelium) and embryonic (blastema) structures mixed with immature mesenchymal cells (stroma) are pathognomonic.

Presence of diffuse anaplasia is associated with poor response to chemotherapy and hence a poorer prognosis (1). This patient did not contain anaplastic elements, hence we expect a good prognosis. Nephroblastomas metastasise only to lymph nodes, lungs and liver. Metastases to other sites are extremely rare.

The prognosis for adults with Wilms' tumour is worse than that for children. Because of the rarity of this neoplasm, no firm treatment guidelines have been established, but a combined approach with surgery, radiotherapy and multi-agent chemotherapy is recommended (4). Surgery includes radical nephrectomy, removal of lymph nodes and assessment of the contralateral kidney for concurrent lesions. The majority of nephroblastomas are treated using therapeutic protocols created by either the International Society of Paediatric Oncology (SIOP) or the Children's Oncology Group (COG). The SIOP protocols advocate preoperative therapy followed by surgical removal. This approach allows for tumour shrinkage prior to resection, yielding a greater frequency and ease of resectability. The COG has long advocated primary resection of tumours followed by therapy that is determined by stage and classification into favorable and unfavorable histology categories. This allows stratification of patients according to pathologic grades. Both SIOP and COG protocols have resulted in similar outcomes (1). Due to its rarity in adults the ideal form of treatment of adult Wilms' tumour is not yet established. Despite the worse prognosis for adult patients with advanced disease, durable complete remissions have been achieved using a multimodal therapy with surgery, chemotherapy and radiotherapy.

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