Pitfalls in the management of infantile renal neoplasia

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This article addresses questions posed by a renal mass in early infancy. Are changes required in the standard approach to a renal mass in this age group? Are diagnostic possibilities transformed enough to warrant a change of approach in a setting like this? What are the 'benign' lesions that require consideration? What is the physician's liability in performing a nephrectomy in a patient with a problem like this? A review of the literature suggests that the vast majority of renal space occupying lesions in children of this age are still Wilms' tumor. Unless diagnostic refinements reach a higher level, the present protocols should be persisted with. *Ann Pediatr Surg* 13:107–109 © 2017 Annals of Pediatric Surgery.

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Introduction

Renal tumors are the second most common abdominal tumors in children, after abdominal neuroblastomas [1]. Wilms' tumor (WT) is by far the commonest renal tumor in children. Other tumors like clear cell sarcoma of the kidney, rhabdoid tumor of the kidney, congenital mesoblastic nephroma, renal cell carcinoma, cystic renal tumor, and angiolipoma of the kidney also occur in significant numbers in this age group, and hence require consideration. The seminal work by Children's Oncology group and Societe Internationale d'oncoligie d'Pediatrique (SIOP) has resulted in clear cut guidelines for the management of renal neoplasia in children. Generally speaking, a clinically detected renal mass should be investigated radiologically and, as per the National Wilms Tumor Study (NWTS) protocol, should be resected and characterized by its histopathological features. This constitutes its local treatment and disease staging [1], both of which determine its further therapy. SIOP protocols generally recommend chemotherapy followed by nephrectomy with surgicopathologic staging at the time of surgery.

However, in the litigious climate of the times, other tumors and nontumor lesions need to be considered in a comprehensive management plan when confronted with space occupying kidney lesions. We present this article based on the case of a young infant with a renal mass, who was treated according to the accepted norms of management of renal tumors and yet the treating team ran into difficulties resulting from its histological diagnosis. The incidence and differential diagnoses of renal neoplasms – benign/malignant – is discussed with reference to cystic renal tumors – for example, cystic nephroma. Xanthogranulomatous pyelonephritis (XGP) in children is reviewed in the context of a non-neoplastic renal mass. Also discussed is the impact of these diagnoses on the professional liability of the physician performing a nephrectomy in such a situation.

Case report

NO was a 5-month-old Saudi girl when referred to us from a secondary care center. She had been suffering from mild abdominal distension, mild diarrhea, and unexplained fever for the last 3 weeks. Her antenatal, natal, and postnatal histories were normal. Examination had not revealed any untoward findings. Blood, urine stool, and cerebrospinal fluid examinations were normal. Ultrasound examination had shown a left renal hypoechoic mass with some hyperechoic structures inside it. Computed tomography (CT) of the abdomen showed a hypodense mass at the lower pole of the left kidney. During her stay in our hospital, several investigations were repeated. Serum ferritin was normal. The C-reactive protein was 3.2 mg/dl. Ultrasound and CT examinations were repeated, which showed a mass, $3 \times 2 \times 3$ cm in size, of heterogeneous density with multiple areas of breakdown (Fig. 1a and b). Perinephric fat was maintained. A perihilar node, 17×10 mm in size, was also seen. Renal vein and inferior vena cava were free of tumor and patent. No metastatic lesions were seen on the chest and skeletal survey. Diagnoses entertained were WT, congenital mesoblastic nephroma, intrarenal neuroblastoma, and renal cell carcinoma. An MRI examination (Fig. 2a) reported a solid, multilocular, neoplastic lesion at the lower pole of the left kidney. Possible diagnoses were the same as for the CT scan examination. As the radiologist had recommended a biopsy before surgical undertaking, an ultrasound-guided fine needle aspiration cytology was carried out. This showed blastemal, epithelial, and stromal cells, which reinforced the argument for WT and a nephrectomy. Accordingly, a left transperitoneal nephrectomy was carried out with dissection of the perihilar and peraortic lymph nodes. Histopathologically, the tumor had large cystic spaces lined with flattened cuboidal epithelium (Fig. 2b), with an infiltrate of inflammatory cells, which agreed with a diagnosis of cystic nephroma. The child was referred to an oncology center for the benefit of a review of her diagnosis and chemotherapy if required. The diagnosis there was adjudged to be XGP and no further treatment was advised.

Discussion

Radiology usually successfully eliminates hydronephroses and multicystic dysplastic kidney, the two commonest

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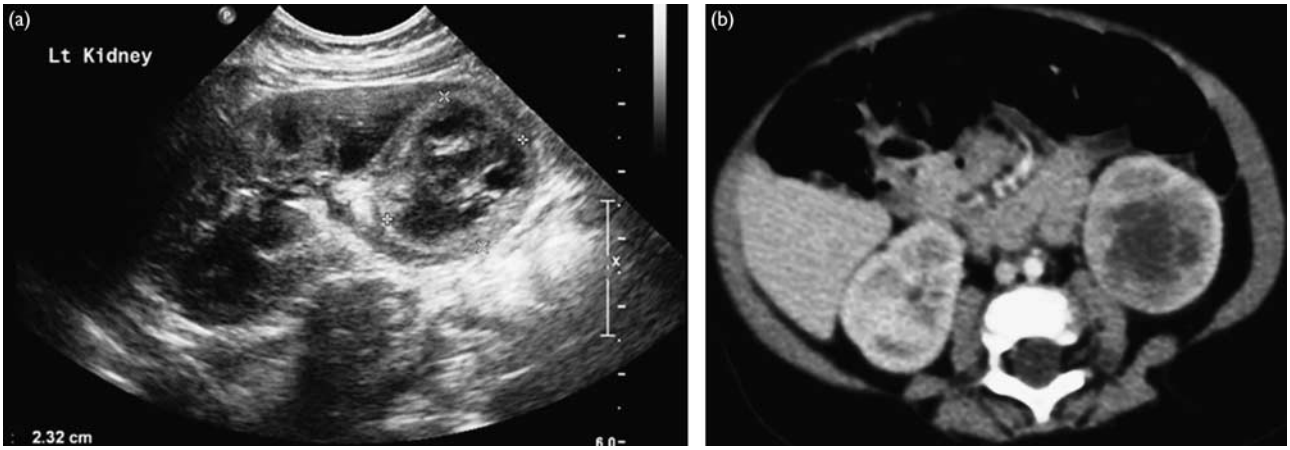
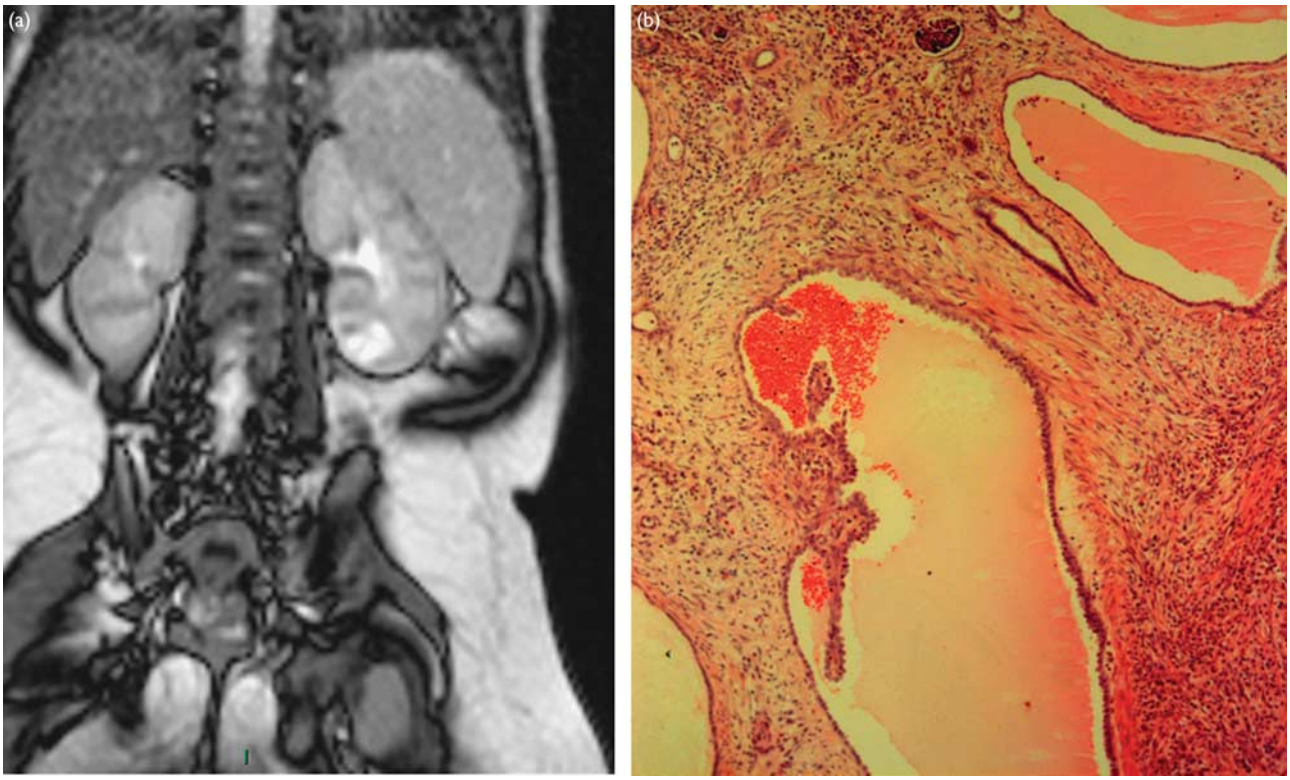
Fig. 1



(a, b) Ultrasonograph of the lesion; longitudinal and transverse views showing the lower pole mass with multiple areas of breakdown.

Fig. 2



(a) MRI, coronal view showing a multilocular 'neoplastic' lesion at the lower pole of the left kidney. (b) Histology of the renal lesion showing large cystic areas lined by cuboidal epithelium diagnosed as cystic nephroma.

causes of renal masses [2]; these affect the entire kidney rather than a sector of it.

In a review of 750 infants, less than 7 months in age, with renal tumors, and drawn from five of the world's largest childhood renal tumor cooperative studies, a large majority (86%) were found to be malignant. Only 6 % of the tumors were non-Wilms' renal tumors [3]. Conversely, in a retrospective review of the radiology of 26 histologically proven non-Wilms' renal tumors, the authors could detect nonspecific attributes of tumors but emphasized the need for histopathologic characterization [4]. The predominance of malignant renal tumors has led to the attitudes adopted in the NWTS and SIOP [5].

The NWTS protocol recommends an upfront nephrectomy on encountering a renal tumor. Pathological staging of the tumor then determines the rest of the treatment. Conversely, the SIOP protocol advocates chemotherapy as the initial treatment to reduce tumor size, which is then followed by surgery [5]. Both approaches initiate

therapy without recourse to preoperative biopsies. In fact, the NWTS avoids preoperative biopsy for fear of tumor seeding and upgrading the pathological stage of the tumor [6]. The UKCCSG adopted preoperative biopsy to accurately determine the diagnosis and institute-appropriate chemotherapy [5,6].

Keeping in view the preponderance of malignant renal tumors, and the absence of other modalities, for example, serological studies [6], that can accurately characterize tumors, nephrectomy remains the only course open to the treating physician.

In the present case, all attempts including an fine needle aspiration cytology were made to accurately establish diagnosis. Though accurate in the UKCCSG studies, its use as a one off diagnostic tool may have limitations as demonstrated in our case.

Partial nephrectomy is being advocated for smaller tumors, to spare as much of a functioning kidney as possible. However, the possibility of a multicentric WT have prevented this from being adopted on a major scale [6].

The present case was diagnosed as a cystic nephroma in our hospital. Multilocular cystic nephromas, as they have been called in the past, are difficult to diagnose preoperatively and nephrectomies have proven satisfactory treatment for them [7]. In the other hospital, the histopathological opinion suggested XGP. XGP, though rare in childhood, may figure in the process of diagnosis because it presents with a renal mass of obscure origin in response to an *Escherichia coli* or *Proteus* spp. urinary tract infection, frequently in the presence of stones or urinary tract obstruction, none of which were seen in this patient. The accompanying fever and other general features of XGP are often seen in association with WT and can add to diagnostic confusion. The virulence of this infection frequently spreads infection to the perinephric fat and even fistulation into other organs. Definitive treatment is achieved by using nephrectomy. In a retrospective review of 17 patients spanning 30 years, only two cases were correctly diagnosed preoperatively [8].

In conclusion, until preoperative diagnostic modalities can determine the nature of a renal lesion more accurately, primary nephrectomy as a modality of treatment is justified.

Conflicts of interest

There are no conflicts of interest.

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