RESIDENT'S CORNER

Metanephric adenofibroma

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A 10-year-old boy underwent a computed tomography (CT) scan for left flank pain following a fall. Imaging demonstrated a 5 cm left upper pole renal mass. Partial nephrectomy revealed metanephric adenofibroma, a benign stromal-epithelial tumor thought to represent a hyperdifferentiated, mature form of Wilms' tumor. We briefly discuss the histopathology and management of this rare tumor.

Key Words: metanephric adenofibroma, Wilms' tumor, pathology, partial nephrectomy, children

Introduction

We report a case of metanephric adenofibroma, a benign metanephric neoplasm. We present the clinical, radiographic, pathologic findings, and management of this rare tumor.

Case report

A 10-year-old otherwise healthy boy presented with left flank pain after a recent fall. Physical examination was unremarkable. A platelet count and hemoglobin were within the normal range, and there was no associated gross or microscopic hematuria. Ultrasonography and contrast-enhanced computerized tomography (CT) of the abdomen and pelvis demonstrated a 5 cm left upper pole solid, homogeneous renal mass, Figure 1. The differential diagnosis was renal cell carcinoma, fat poor angiomyolipoma, lymphoma, sarcoma, mesoblastic nephroma, Wilms' tumor or Wilms' spectrum benign variant (mesenchymal metanephric stromal tumor, epithelial metanephric adenoma, mixed metanephric adenoma). The index of suspicion for Wilms' tumor was low, given the patient's age and benign imaging characteristics. After appropriate preoperative counseling, including the potential of off-protocol treatment in the instance that the final pathology was Wilms' tumor, a decision was made to proceed with left partial nephrectomy, which was performed through a subcostal incision. Intraoperative frozen section indicated a negative resection margin. Gross examination revealed an encapsulated ovoid and lobular tan-pink soft tissue mass, Figure 2a and 2b. Final histopathologic evaluation was that of a metanephric adenofibroma.

Discussion

Metanephric adenofibroma, formerly designated nephrogenic adenofibroma, is a rare, biphasic, benign stromal-epithelial tumor. This tumor is classified among the metanephric neoplasms, which also include metanephric adenoma (a purely epithelial lesion) and metanephric stromal tumor (a purely stromal lesion). Metanephric neoplasms are thought to represent the most hyperdifferentiated, mature form of Wilms' tumor on the benign end of the developmental spectrum.[1,2] These tumors are composed of varying proportions of mitotically inactive epithelial and stromal tissue. While this tumor has been described in individuals in the fourth decade of life, the majority are diagnosed in children and teenagers (mean age 10.2 years).[1-3] Patients commonly present with hematuria, as the renal medullary origin facilitates collecting system invasion.[1] They can achieve substantial size, thus risking confusion with malignant renal tumors as demonstrated by

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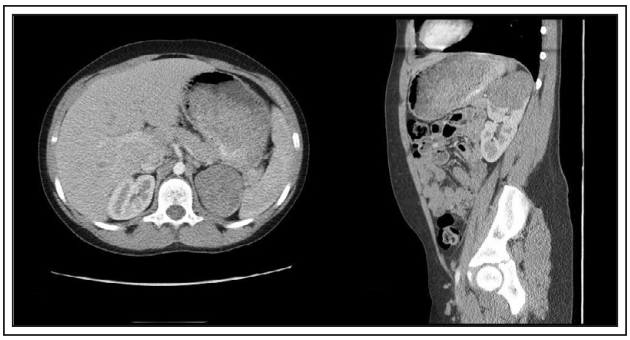

Figure 1. Contrast-enhanced CT shows a well-defined 5 cm solid mass arising from the upper pole of the left kidney. The renal vessels were patent bilaterally and no lymphadenopathy was identified.

Piotrowski et al, who described robotic assisted partial nephrectomy for a 19 cm metanephric adenofibroma.[4]

Metanephric adenofibroma is usually a solitary tumor with indistinct borders centered in the renal medulla. Grossly, the lesions are partially cystic or nodular and have a firm consistency with tan, white-grey, or yellow coloration. Areas of hemorrhage and necrosis have been observed with concomitant Wilms' tumor.[3,5] The epithelial nodules are unencapsulated and composed of tightly packed tubular, tubulopapillary, or papillary structures. Psammoma bodies—eosinophilic extracellular concentric calcified structures seen in a variety of papillary neoplasms—are a common microscopic finding.[2,5] Some lesions have increased mitotic activity (> 5 mitoses/20 hpf) and have been subclassified as "metanephric adenofibroma with epithelial mitoses". The spindled stromal component features angiodysplasia, concentric "onion skin" peritubular growth pattern, and glial differentiation.[3,5] At this point in time, no immunohistochemical profile exists to characterize metanephric neoplastic lesions, although the epithelial component is typically positive for cytokeratin (AE1/3 or CAM 5.2) and WT-1. The stromal component is frequently positive for vimentin and CD34 with variable expression of CD117 (c-kit proto-oncogene product).[1,6] EMA, actin and desmin stains are negative.[5] Microsatellite allelotyping has suggested imbalance at chromosome 2p13 in over 50% of metanephric adenomas, but is not described for the biphasic metanephric adenofibroma.[1]

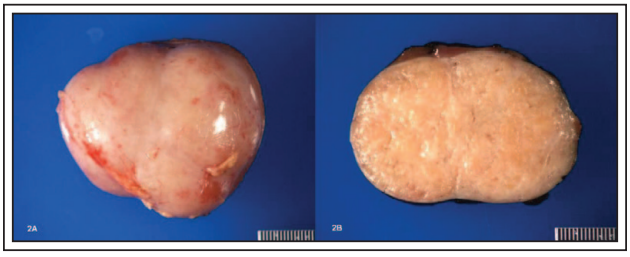

Figure 2. The tumor consisted of a 62 gr ovoid, lobular tan-pink soft tissue mass measuring 5.8 cm x 4.7 cm x 4.0 cm. The outer surface was smooth and glistening with a small area of fibroadipose connective tissue **(a)**. Cut sections revealed an encapsulated tumor that had a granular, tan-pink and fibrous appearance **(b)**.

Some patients with metanephric adenofibroma are given an initial diagnosis of Wilms' tumor, mandating adjuvant chemotherapy, which makes it difficult to assess the inherent biologic potential of the tumor.[1-3] The morphologic features and low mitotic index, however, suggest a benign course. To date, no recurrences following surgical resection by radical or partial nephrectomy have been reported in the literature. It is the current recommendation to perform complete excision of metanephric neoplasms, including adenomas, adenofibromas, and stromal tumors, to eliminate the risk of underlying concurrent malignancy or malignant transformation of residual microscopic disease. It is the opinion of the authors that partial nephrectomy should be considered when technically feasible. Very rare cases of metastatic metanephric adenoma and adenosarcoma at presentation have been reported and long term behavior is unknown due to lack of follow up in the literature.[6,7] Postoperatively, this patient was discussed at our multi-disciplinary tumor board conference. Based on the aforementioned data that postoperative recurrence is unreported, the consensus opinion was for no further postoperative imaging or testing beyond routine visits with the child's pediatrician. □

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