Radiology Case Reports 13 (2018) 610–613

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://Elsevier.com/locate/radcr

Pediatric

An "accidental" discovery: Incidentally found metanephric adenofibroma in a 5-year-old male trauma patient

Priya G. Sharma MD[a,*], *Ashwini K. Esnakula MD*[b], *Dhanashree A. Rajderkar MD*[a]

[a] *Department of Radiology, University of Florida College of Medicine, 1600 SW Archer Road, PO Box 100374, Gainesville, FL 32608-0374*

[b] *Division of Surgical Pathology, Department of Pathology, University of Florida Gainesville, Gainesville, FL, USA*

ARTICLE INFO

Article history:
Received 22 February 2018
Accepted 26 February 2018
Available online

Keywords:
Pediatric renal mass
Wilms
Benign mass

ABSTRACT

Metanephric adenofibroma is a rare pediatric renal tumor, which should be considered in cases of solid renal lesions that mimic Wilms tumor on both imaging and histology. We present a case of an incidentally found left renal lesion on a trauma computed tomography scan in a 5-year-old male patient. The patient underwent total nephrectomy, and the diagnosis of metanephric adenofibroma was made on histology. Radiologists should consider this entity in the differential for an incidentally found solitary renal mass in a pediatric patient. Prompting the pathologic diagnosis of this entity can spare patients from unnecessary chemotherapy and allow for nephron-sparing surgery.

Introduction

Metanephric neoplasms are rare renal neoplasms identified in both pediatric and adult populations. These neoplasms consist of metanephric adenoma, metanephric stromal tumor, and metanephric adenofibroma (MAF). MAF consists of both epithelial and stromal cells and has even been reported to merge with the morphology of Wilms tumor [1]. Less than 100 of these cases have been reported in the literature, appearing more often within pathology literature [2]. The imaging appearance of MAF is nonspecific, with the tumor often resembling the Wilms tumor. However, the management of the lesions is quite different, and correctly diagnosing MAF may spare the patient from receiving toxic chemotherapy. Therefore, MAF remains an important pathology for the radiologist to consider when reporting solid renal lesions in the pediatric population.

Case report

An otherwise healthy 5-year-old male patient, who was a restrained passenger in a high-speed motor vehicle collision,

Competing Interests: The authors have declared that no competing interests exist.
Funding: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.
Author Contributions: Priya G. Sharma, MD: conception, drafting of the article, and intellectual radiological content. Ashwini Esnakula, MD: drafting of the article and intellectual pathology content. Dhanashree A. Rajderkar, MD: conception, critical revision, and final approval of version to be submitted.

* *Corresponding author.*
E-mail address: sharpg@radiology.ufl.edu (P.G. Sharma).

https://doi.org/10.1016/j.radcr.2018.02.026

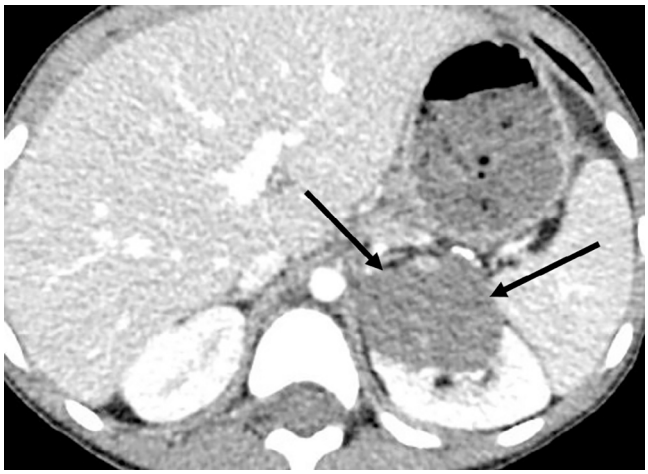

Fig. 1 – Axial contrast-enhanced computed tomography imaging demonstrating a homogenous solid mass with no areas of calcification or necrosis (black arrows) in the superior pole of the left kidney.

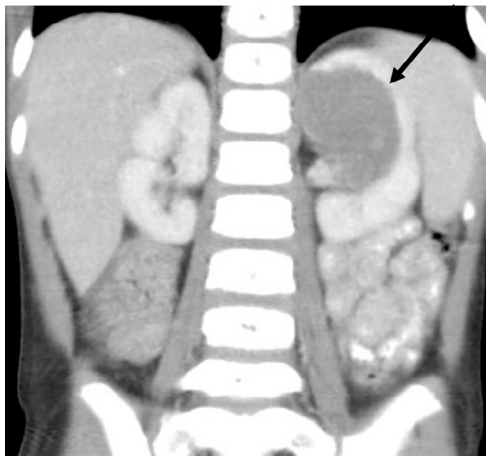

Fig. 2 – Coronal contrast-enhanced computed tomography image demonstrating a solid hypodense mass (black arrow) arising from the superior pole of the left kidney.

presented to the pediatric emergency room as a trauma transfer from the regional medical center for pediatric surgery evaluation. Contrast-enhanced computed tomography (CT) scan from outside the hospital (dose-length product 111.70 mGy·cm) reported a 3.8-cm mass arising from the anterior aspect of the left kidney, suggestive of a neoplastic process such as Wilms tumor. Review of the CT scan showed a low-attenuating lesion without calcification or necrosis. There was no invasion of the renal vein or retroperitoneal adenopathy. The kidneys showed no additional lesions to suggest nephrogenic rests. The perinephric fat planes were preserved. No metastatic lesions were identified in the abdomen or the pelvis. There were no traumatic findings otherwise (Figs. 1-3). Ultrasound was performed when the patient arrived at our institution for confirmation. Ultrasound revealed a $5.4 \times 3.3 \times 3.3$ cm solid iso- to hypoechoic mass with mild internal vascularity emanating from the left kidney and therefore concerning for Wilms tumor (Figs. 4 and 5). There was no retroperitoneal adenopathy or invasion of the renal vein. Metastatic workup with CT of the chest was negative.

After discussion of management options with the patient's parents, decision was made to undergo left nephrectomy. Because of potential for tumor upstaging, biopsy of this unilateral tumor was not recommended. Given the patient's history of trauma and "seatbelt" sign on presentation, an urgent same-day exploratory laparotomy was performed. Surgery was complicated by the presence of intraperitoneal blood and desvascularized small intestine at the distal ileum with deserosalized and perforated intestine. Small bowel resection was performed with a primary anastomosis. A complete left nephrectomy was performed with no tumor spillage. Regional periaortic lymph nodes were dissected.

Intraoperative frozen section demonstrated spindle cells, and a definitive diagnosis was deferred by pathology at the time.

Pathology revealed a $4.5 \times 4.5 \times 4.0$ cm firm well-circumscribed mass limited to the right renal parenchyma (Fig. 6A). The mass had a homogenous white-to-tan cut surface without any areas of hemorrhage or necrosis. Microscopic images demonstrated a biphasic tumor with stromal and epithelial components (Fig. 6B). The stromal component was predominantly composed of spindle to stellate cells with thin tapered hyperchromatic nuclei. The stromal cellularity varied from hypocellular myxoid areas to cellular fibroblastic areas (Fig. 6C). The stromal cells were diffusely positive for CD34 by immunohistochemistry. The epithelial component was limited and was mostly identified in the subcapsular areas as unencapsulated nodules. These epithelial nodules consisted of a cellular inactive embryonal-type epithelium composed of uniform, small cuboidal cells with small hyperchromatic nuclei and scant cytoplasm forming small tubules and blunt short

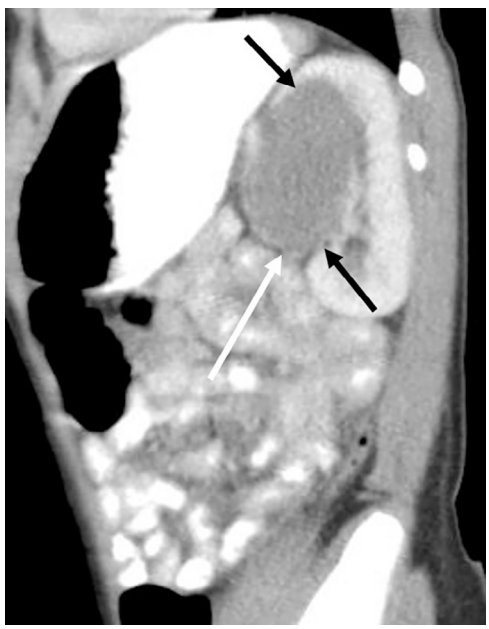

Fig. 3 – Sagittal contrast-enhanced computed tomography image demonstrating the "claw sign" (black arrows) consistent with mass (white arrow) arising from the superior pole of the left kidney.

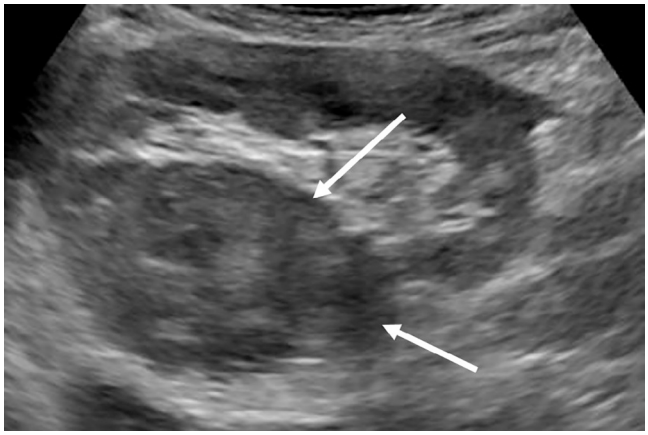

Fig. 4 – Ultrasound demonstrating a well-defined iso- to hypoechoic (to the renal cortex) solid mass (white arrows) emanating from the superior pole of the left kidney.

papillae resembling glomeruli (Fig. 6D). Overall, these histologic findings were consistent with a MAF.

After a short course of intravenous antibiotics for intermittent postoperative fevers, after remaining afebrile, the patient transitioned to oral antibiotics and was discharged home.

A single follow-up retroperitoneal ultrasound 6 months later, which was negative for recurrence, confirmed a normal right kidney. Given the pathology, no further follow-up was felt necessary.

Discussion

MAF is a very rare renal tumor found in both children and adults. The tumor falls into the larger category of metanephric tumors. Histologically, metanephric tumors contain epithelial cells, stromal cells, or both and are respectively referred to as metanephric adenoma, metanephric stromal tumor, or MAF.

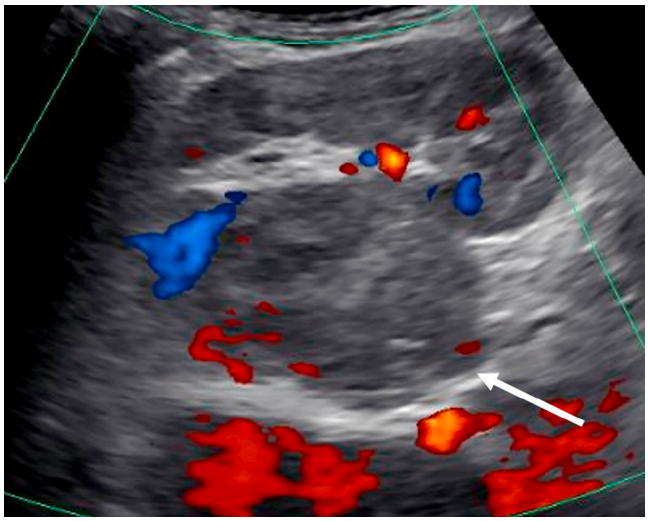

Fig. 5 – Doppler ultrasound showing mild vascularity to the renal mass (white arrow).

Metanephric adenoma is the most common of the lesions and typically affects adult women [3]. Metanephric stromal tumors, on the other hand, typically present in young children with a mean age of presentation of 2 years [4]. MAF was originally termed "nephrogenic adenofibroma" and was described in 1992 by Hennigar and Beckwith [5]. Since that time, review of the literature shows that less than 30 cases have been reported in children. Although MAF is considered benign, it is difficult to distinguish from malignant lesions by imaging characteristics alone. Definitive diagnosis is typically made after surgical resection by radical or partial nephrectomy.

As in our case, more than half the reported cases of MAF have been found in asymptomatic patients [2]. However, patients can present with gross hematuria when a centrally located lesion penetrates the collecting system. Clinical presentation also includes pain, hypertension, and polycythemia owing to the tumor's production of erythropoietin [1].

Solid renal tumors in the pediatric population are thought to be Wilms tumor. However, the differential for homogenous solitary solid renal lesions in the pediatric population should also include renal cell carcinoma, clear cell sarcoma, and lymphoma in the appropriate clinical setting. Aggressive features such as vascular invasion and metastatic or bilateral lesions have not been reported features of MAF, and if these imaging features are seen, alternate diagnoses should be suggested. Imaging provides no guidance in the determination of an early Wilms tumor vs the MAF. In fact, the 2 pathologies tend to share many imaging features. On CT, the lesion appears homogenously hypovascular to the renal parenchyma. Lesions can be predominantly cystic and associated with calcifications [6,7]. Wilms tumor also tends to be hypovascular to the renal parenchyma when diagnosed early. Calcifications are seen in only 20% of Wilms tumors. Unfortunately, calcifications are also seen in pediatric renal sarcomatous lesions. Echogenicity of the MAF varies on ultrasound [8]. Case reports describe no benefit to the use of magnetic resonance imaging in the characterization of this lesion. Again, this lesion shares imaging features seen in other renal neoplasms. On T1- and T2-weighted imaging, this lesion remains hypointense. After the administration of gadolinium, this lesion demonstrates a lower signal intensity than the surrounding parenchyma [2]. Imaging features of a solitary mass without local, regional, or distant metastases should prompt the radiologist to consider MAF in an asymptomatic patient.

Biopsy of unilateral renal lesions is not recommended by the Children's Oncology Group protocol as this leads to tumor upstaging [7]. Furthermore, fine-needle aspiration to effectively diagnose MAF has not been demonstrated in the literature [6]. In terms of surgical approach, if MAF is confirmed by initial biopsy or frozen section at the time of surgery, because of the tumor's benign course, nephron-sparing surgery is advocated [9]. Adjuvant chemotherapy is not necessary [10].

Rarely, MAF are associated with malignant neoplasms such as nephroblastoma or papillary renal cell carcinoma [1]. There was no histologic evidence of a nephroblastoma or a papillary renal cell carcinoma in this current case. Recent studies show that metanephric family tumors including MAF frequently show B-rapidly accelerated fibrosarcoma (*BRAF*) V600E mutations [4,11,12], which is not identified in nephroblastomas. In our case, immunohistochemistry for *BRAF* V600E mutation showed diffuse moderate to strong positivity in the epithelial

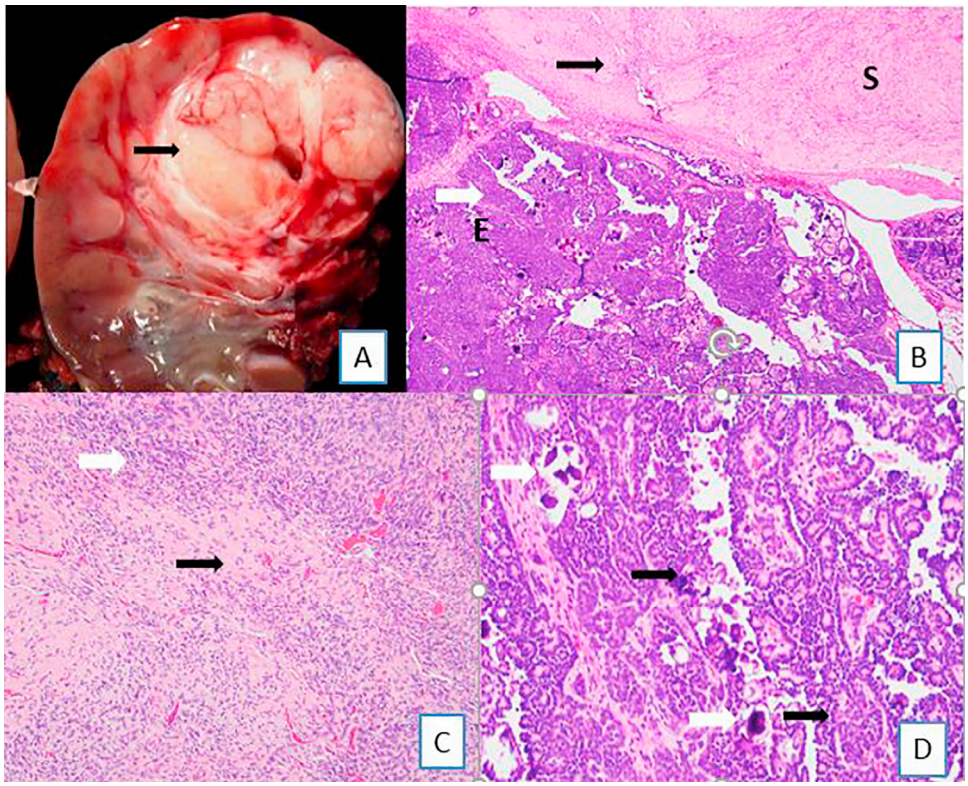

Fig. 6 – (A) Right kidney with a tan homogenous mass in the upper pole S (black arrow). (B) Biphasic tumor with S and E components (H&E 40×). (C) S component consistent with metanephric S tumor with hypocellular (dark black arrow) and hypercellular E (white arrow) areas composed of spindle to stellate cells. (D) E component consistent with metanephric adenoma composed of a cellular inactive embryonal-type epithelium S (black arrows) and frequent psammomatous calcifications E (white arrows). H&E, hematoxylin and eosin; E, epithelial; S, stromal.

component and patchy moderate positivity in the stromal component. Targeted next-generation sequencing confirmed the presence of *BRAF* V600E (c.1799T>A) mutation in the tumor. In addition, *PTEN* c.737C>T p.P246L was also identified. To our knowledge, *PTEN* mutations have not been reported in metanephric tumors. Cytogenetic studies showed normal karyotype and no clonal cytogenetic abnormalities were identified. These findings further supported the diagnosis of a MAF.

MAF is an extremely rare tumor that should be considered in the differential for a non–aggressive appearing solitary pediatric renal mass presenting in an asymptomatic patient.

REFERENCES

[1] Arroyo MR, Green DM, Perlman EJ, Beckwith JB, Argani P. The spectrum of metanephric adenofibroma and related lesions: clinicopathologic study of 25 cases from the National Wilms Tumor Study Group Pathology Center. Am J Surg Pathol 2001;25(4):433–44.

[2] Amodio JB, Shapiro E, Pinkney L, Rivera R, Strubel N, Douglas D, et al. Metanephric adenoma in an 8-year-old child: case report and review of the literature. J Pediatr Surg 2005;40(5):e25–8.

[3] Davis CJ Jr, Barton JH, Sesterhenn IA, Mostofi FK. Metanephric adenoma: clinicopathological study of fifty patients. Am J Surg Pathol 1995;19(10):1101–14.

[4] Argani P, Beckwith JB. Metanephric stromal tumor: report of 31 cases of a distinctive pediatric renal neoplasm. Am J Surg Pathol 2000;24(7):917–26.

[5] Hennigar RA, Beckwith JB. Nephrogenic adenofibroma. A novel kidney tumor of young people. Am J Surg Pathol 1992;16(4):325–34.

[6] Bastide C, Rambeaud JJ, Bach AM, Russo P. Metanephric adenoma of the kidney: clinical and radiological study of nine cases. BJU Int 2009;103:1544–8.

[7] Saltzman AF, Carrasco A Jr, Maccini MA, Caldwell BT, Treece AL, Cost NG. Large cystic metanephric adenoma in a 15-year-old girl. Urology 2017;101:147–50. doi:10.1016/j.urology.2016.12.019 [Epub 2016 Dec 21].

[8] Kacar A, Azili MN, Cihan BS, Demir HA, Tiryaki HT, Argani P. Metanephric stromal tumor: a challenging diagnostic entity in children. J Pediatr Surg 2011;46(12):e7–10. doi:10.1016/j.jpedsurg.2011.08.023.

[9] Yao DW, Qu F, Hu SW, Zheng JY, Wang JM, Zhu XY, et al. Metanephric adenofibroma in a 10-year-old boy: report of a case and review of the literature. Int J Clin Exp Pathol 2015;8(3):3250–6.

[10] Raj P, Khanolkar A, Sarin YK. Metanephric adenofibroma masquerading as Wilms' tumor. APSP J Case Rep 2016;7(5):37.

[11] Chami R, Yin M, Marrano P, Teerapakpinyo C, Shuangshoti S, Thorner PS. BRAF mutations in pediatric metanephric tumors. Hum Pathol 2015;46:1153–61.

[12] Argani P, Lee J, Netto GJ, Zheng G, Tseh-Lin M, Park BH. Frequent BRAF V600E mutations in metanephric stromal tumor. Am J Surg Pathol 2016;40(5):719–22.