# Large Cystic Metanephric Adenoma in a 15-Year-Old Girl



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Metanephric adenoma is a rare pediatric renal tumor, generally considered to be benign. It can be difficult to distinguish from Wilms tumor and renal cell carcinoma based on imaging alone, and even may be difficult on histopathologic analysis. We present a case of a large cystic metanephric adenoma managed with surgical resection. This case highlights the difficulty in managing cystic renal lesions in children and adolescents as there is a paucity of data on the radiologic and pathologic correlation in such patients. UROLOGY 101: 147–150, 2017. © 2016 Elsevier Inc.

etanephric adenoma (MA) is a rare renal tumor (0.2% of adult renal masses<sup>1</sup>) arising from the renal medulla.<sup>2</sup> These tumors are generally reported in middle-aged women,<sup>1</sup> with around 20 case reports in the pediatric literature.<sup>3</sup> It is considered a benign tumor, although there have been case reports of regional lymph node involvement<sup>2,4</sup> (7- and 11-year-old female patients) as well as bone metastases<sup>5</sup> (32-year-old woman). Imaging typically reveals a solid, well-circumscribed lesion.<sup>1</sup> Unfortunately, it cannot be definitively differentiated from Wilms tumor (WT) or renal cell carcinoma (RCC) based on imaging characteristics alone, and it requires histologic evaluation.<sup>1</sup> Thus, definitive diagnosis is not typically made until after surgical resection by either radical or, when feasible, partial nephrectomy.<sup>6</sup> We present a case of a large cystic MA managed with surgical resection via partial nephrectomy.

## **CASE PRESENTATION**

An asymptomatic 15-year-old adolescent girl was referred to pediatric urology after physical examination revealed a palpable mass of the right upper quadrant. Medical and surgical histories were negative. Laboratory evaluation was normal. Computed tomography (CT) of the abdomen and pelvis was obtained, which revealed a 14 cm complex cystic lesion of the anterior lower pole of the right kidney (Fig. 1A-E). There were associated calcifications; therefore, this was graded as a Bosniak 2F cyst. Metastatic workup with CT of the chest was negative. After discussion of management options, the patient and her parents elected to pursue surgical extirpation. She was enrolled in the Children's Oncology Group renal tumor biology and banking study (AREN 03B2).

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Per protocol, the patient underwent a right open partial nephrectomy with regional lymph node dissection (Fig. 2A,B). A minimally invasive approach was discussed and considered but ultimately not used owing to concern for this representing a cystic malignancy and a potential risk for rupture if mishandled. Also, we took into consideration the size of the lesion, which would necessitate a large incision for extraction. Direct, manual renal parenchymal clamping was used for hemostatic control, and thus no global renal ischemia was induced. A complete resection was accomplished with a grossly negative surgical margin and no tumor rupture or spillage. Intraoperative frozen section revealed a benign cystic process, but a definitive diagnosis was deferred by pathology at that time. Blood loss was 200 mL and operative time was 3.5 hours. Her hospital course was uneventful.

Pathologic evaluation revealed a 16 cm cystic MA with 14 benign lymph nodes. The tumor was encapsulated and the cyst lining had scant yellow-tan excrescences that corresponded to areas of cellularity (Fig. 3A,B). The tumor cells were arranged in a vaguely tubular and papillary architecture with bland, round nuclei and pale eosinophilic cytoplasm. Immunohistochemical staining was positive for nuclear WT-1 (Fig. 1C) and negative for CD57. Additional areas of cyst wall calcification and ossification were present.

The patient underwent renal ultrasound 6 months postoperatively, which was negative for recurrence. Her imaging will be repeated in 1 year.

### DISCUSSION

MA (pure epithelial) metanephric stromal tumor (pure stromal) and metanephric adenofibroma (mixed)<sup>7,8</sup> belong to a group of 3 renal metanephric tumors with a generally benign course. MA was first described by Bove et al in 1979,<sup>9</sup> with most cases being reported in the pathology literature. Although considered benign, reports of metastases have been published.<sup>2,4,5</sup>

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**Figure 1.** Preoperative computed tomographic scan showing a large cystic lesion arising from the anterior lower pole of the right kidney. Noncontrasted phase images showing peripheral calcifications (**A**,**B**), nephrogenic phase showing nearness to the hilum and collecting system (**C**,**D**), and excretory phase showing nearness to the collecting system (**E**).

Approximately half of these lesions are detected incidentally<sup>6</sup>; however, some are associated with symptomatology. MA is the renal tumor most commonly associated with polycythemia (12%),<sup>1</sup> possibly owing to tumor production of erythropoietin and other cytokines.<sup>6</sup> A case series from the Memorial Sloan Kettering Cancer Center notes a possible association with maternal breast cancer (3 of 8 patients), although this has not been consistently reported in the literature.<sup>6</sup> Despite the female predominance, there have been no reports of these tumors having estrogen or progesterone receptors.<sup>6</sup>

On cross-sectional imaging, these lesions are usually solid and hypovascular.<sup>6</sup> They can be associated with cysts<sup>10</sup> and calcifications.<sup>6,10</sup> On Doppler ultrasonography, the echogenicity of the lesion can vary,<sup>1</sup> but there is usually little blood flow seen.<sup>6</sup> Noncontrasted CT scans may show calcifications, whereas contrast-enhanced CT scans confirm the tumors to be solid and homogenous with only mild enhancement.<sup>1</sup> These tumors have lower attenuation after contrast administration than does the renal parenchyma.<sup>6</sup> Limited data are available on the appearance of this tumor on magnetic resonance imaging scans. In case reports, these lesions are usually hypointense on non–gadolinium-enhanced T1 and T2 imaging, and again have lower intensity signal than does the surrounding renal parenchyma,<sup>6</sup> adding little to the ability to preoperatively diagnose this tumor. These imaging characteristics can be explained by the low vascularity of the tumors, which has been confirmed with angiography studies.<sup>6</sup> There is only a single report of a completely cystic mass as in our case.<sup>11</sup> The variable and nonspecific radiographic findings of these tumors make it difficult to distinguish MA from WT or RCC; histopathology is needed for definitive diagnosis.<sup>12</sup>

Our case was a largely cystic lesion. In adults, the Bosniak classification system would give guidance as to the likelihood of malignancy and suggest therapeutic options,<sup>13</sup> but this system has not been validated in children. There are no clear guidelines on how to handle complex cystic lesions in the pediatric population.

Biopsy has been advocated by some, with conflicting reports as to the success of fine needle aspiration to effectively establish an MA diagnosis,<sup>6</sup> because this lesion can be very difficult to differentiate from other malignant tumors. Biopsy of unilateral tumors is not recommended under current Children's Oncology Group protocols as this results in tumor upstaging. Additionally, in cystic cases such as ours, the nondiagnostic rate of renal biopsy is likely to be higher than with a solid lesion.<sup>14</sup> If MA is diagnosed



**Figure 2.** Intraoperative photos showing cystic lesion after complete mobilization **(A)**; remaining renal parenchyma after resection and regional lymphadenectomy **(B)**. (Color version available online.)



Figure 3. Pathologic specimen (A), gross cyst wall (B), and WT-1 staining (C). (Color version available online.)

preoperatively, consideration of observation vs surgical intervention can be discussed. Because the natural history of these tumors is largely unknown, most reports support radical or partial nephrectomy for these lesions. There have been few reports of observation for these lesions, albeit with short follow-up.<sup>6</sup> The natural history of this lesion is not known as most tumors are resected on diagnosis. There is a single case report of 5 years of observation of a child with an MA, which showed exponential growth of the tumor before excision.<sup>15</sup> Postresection follow-up is not well defined.

On pathologic examination, these tumors usually comprise epithelial elements arranged in tubules. The tumor, although well circumscribed,<sup>12</sup> typically has a discontinuous capsule if any capsule at all.<sup>6</sup> There are often associated psammoma bodies.<sup>6</sup> Immunohistochemical staining is variable, although most tumors are positive for WT-1 and CD57, unlike our case, which was negative for CD57. This lesion can often be confused with an epithelial WT variant or papillary RCC. Fluorescence in situ hybridization can be useful to differentiate MA from papillary RCC, the latter generally showing chromosome 7, 17, and Y aneuploidy.<sup>3</sup> Although atypical features can be present, these do not predict the risk of metastases as abnormal pathologic features have been reported in both metastatic and non-metastatic cases.<sup>8</sup> Similarly, 2 cases of pediatric regional lymph node metastases were reported in patients without cytologic atypia.<sup>10</sup>

#### CONCLUSION

MA is a rare renal tumor that can be difficult to distinguish from other pediatric renal tumors. Although it is considered benign, metastatic lesions have been reported. Because MA is difficult to distinguish from WT and RCC, and its natural history is unknown, we advocate for resection and subsequent surveillance. This case highlights the difficulty in managing cystic renal lesions in children and adolescents as there is a paucity of data on the radiologic and pathologic correlation, demonstrating the need for future consensus guidelines.

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