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Metanephric Adenoma with cystic changes- An uncommon presentation of a rare tumor in a young adult

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ABSTRACT

Metanephric adenoma (MA) is a rare benign neoplasm of the kidney that is usually asymptomatic and incidentally diagnosed. MA usually present as a solid mass; however, a cystic presentation has been reported. The main differential diagnosis of MA is the epithelial predominant Wilms tumor (e-WT) and the solid variant of papillary renal cell carcinoma (pRCC). The presence of the BRAF gene mutation has recently been reported in 85% of MA, and less than 10% of cases of MA do not express this specific gene mutation. Herein we report a 22-year-old man who presented with back pain and abdominal discomfort with a renal mass on the computed tomographic scan. The diagnosis of metanephric adenoma was confirmed histopathologically. In our case, the tumor presented as a solid and cystic mass hence mimicking a papillary renal cell carcinoma. The VE1 protein, which correlates with BRAF gene mutation, did not show any significant expression. We want to highlight that MA can present as a cystic lesion that should be taken into account to avoid unnecessary radical nephrectomy. Also, we demonstrated that a subset of MA might not harbor the BRAF gene and, they are classified as the BRAF wild type MA.

Keywords
Proto-Oncogene Proteins B-raf; Kidney Neoplasm, Adenoma

INTRODUCTION

Metanephric adenoma (MA) of the kidney is rare and benign neoplasms. They are usually incidentally diagnosed due to its asymptomatic course and present as a solid mass but may be rarely cystic.[1] About 85% of MA is associated with the mutation of the BRAF oncogene.[2] Less than 200 cases have been reported worldwide in the English literature.[3] It is reported that MA accounts for approximately 0.2% of the adult renal epithelial neoplasms.[4] It generally occurs in adults and has an excellent prognosis.[5] The origin of these neoplasms is not entirely known; however, they are thought to be associated with Wilms tumor.[6] These neoplasms may mimic other renal tumors such as the epithelial predominant Wilms tumor (e-WT) in children and the solid variant of the papillary renal cell carcinoma (pRCC) in adults.[7]

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CASE REPORT

A 22-year-old man presented with back pain and abdominal discomfort. He had no history of hematuria or dysuria. His physical examination and laboratory tests were normal. Computed tomographic scan of the pelvis revealed a complex solid cystic lesion in the mid and lower pole of the right kidney with few foci of punctate and linear calcification, measuring 9.7 x 8.2 x 7.7 cm. (Figure 1A and 1B).

A provisional diagnosis of RCC was established. A right nephrectomy was performed, which showed a solid cystic mass occupying the lower pole and part of the middle pole of the right kidney with 8.5x6.5x6 cm. Grossly, the tumor showed a variegated appearance with areas of necrosis and hemorrhage. (Figure 2A and 2B). The microscopy revealed a well-circumscribed, un-encapsulated pushing mass, composed of monotonous, closely packed tubules, acini along with few solid components. The tumor cells lining the tubules and acini were small, uniform and, round primitive-appearing cells. The intervening

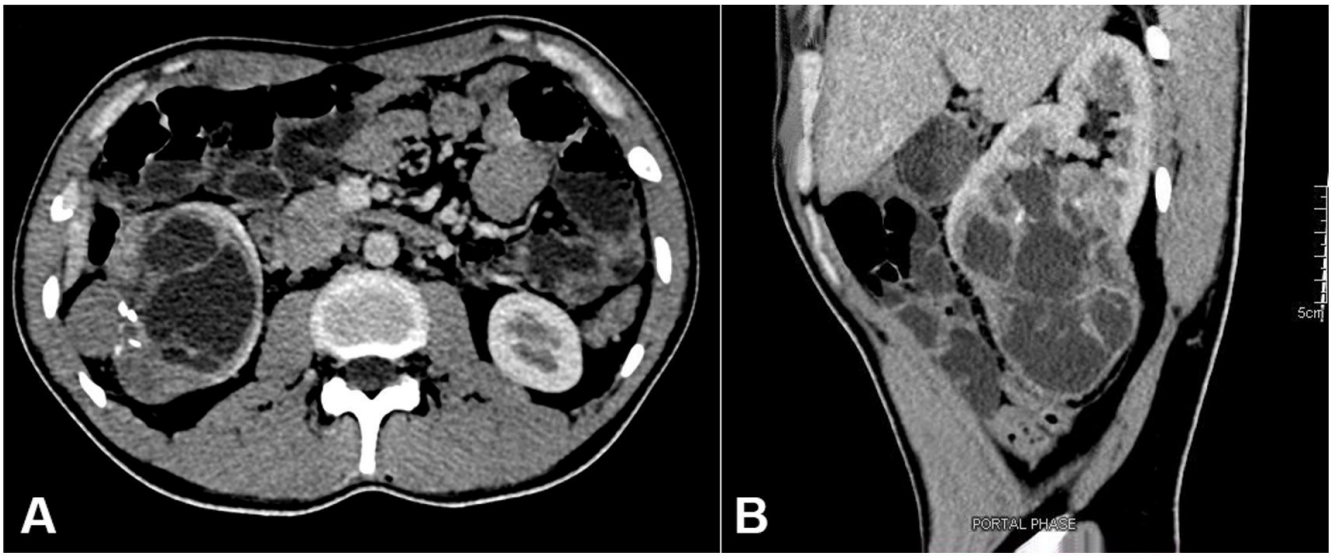

Figure 1. Abdominal contrasted CT showing in **A** – axial plane - and in **B** – sagittal plane – a 9.7 X 8.2 X 7.7 cm complex solid and cystic mass in the mid and lower pole of the right kidney with few foci of punctate and linear calcification.

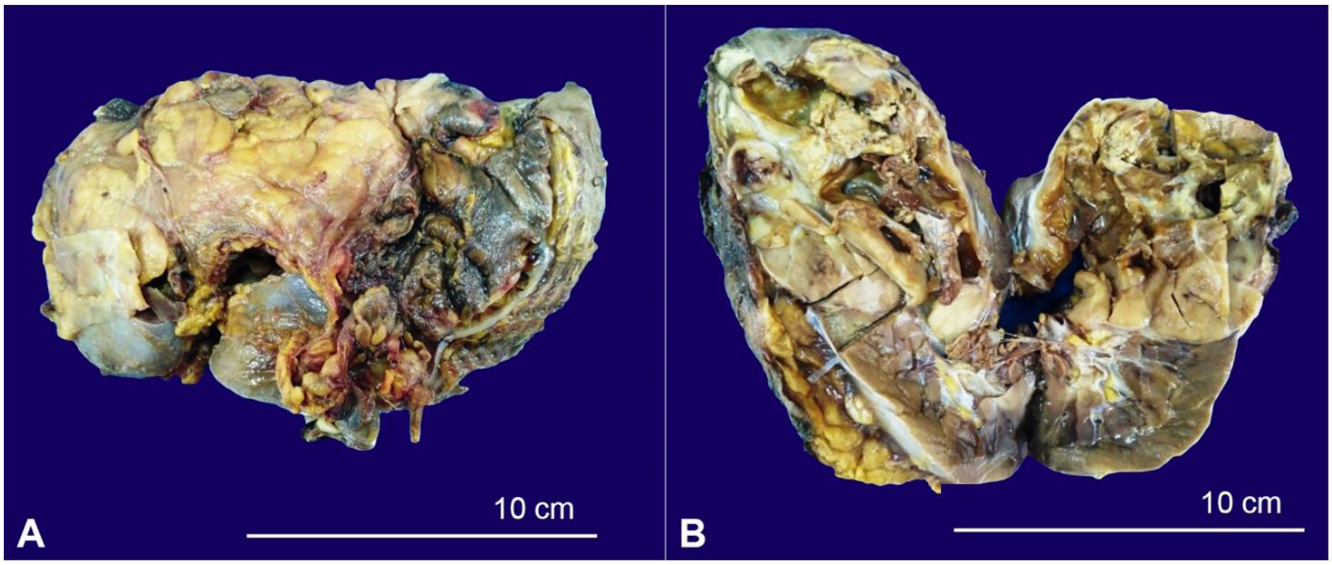

Figure 2. Gross view of the surgical specimen showing in **A** and **B** a solid, and cystic mass occupying two-thirds of the renal parenchyma.

stroma was hyalinized to loosely edematous and showed within it numerous interspersed fine delicate vessels. Psammoma bodies were also seen throughout the tumor. Mitosis, however, was rare.

Large areas of hemorrhage, necrosis and cystic degeneration were noted. However, neither lymphovascular nor perineural invasion was seen. (Figure 3A, 3B, 3C & 3D). Immunohistochemical stains revealed strong nuclear positivity for WT1 and no expression of CK7, AMACR, and CD10. (Figure 4A, 4B & 4C). Ki-67 proliferative index was 1%. The microscopic and immunohistochemical staining results were consistent with the diagnosis of metanephric adenoma. Negative staining for CK7, AMACR, and CD10 rules out pRCC while a low Ki-67 proliferative index rules out Wilms tumor. VE1 protein expression, which correlates with BRAF V600E gene mutation, was found to be negative on immunohistochemistry (Figure 4D).

DISCUSSION

Bove et al. described the term 'Metanephric adenoma' which originates from the primitive epithelium of the proximal nephron.[3] MA is a rare benign renal neoplasm that may clinically and histopathologically mimic other malignant renal tumors.[4] This tumor occurs at a wide age distribution but generally afflicts middle-aged individuals with a female predominance (2:1).[1] In children, MA overlaps in

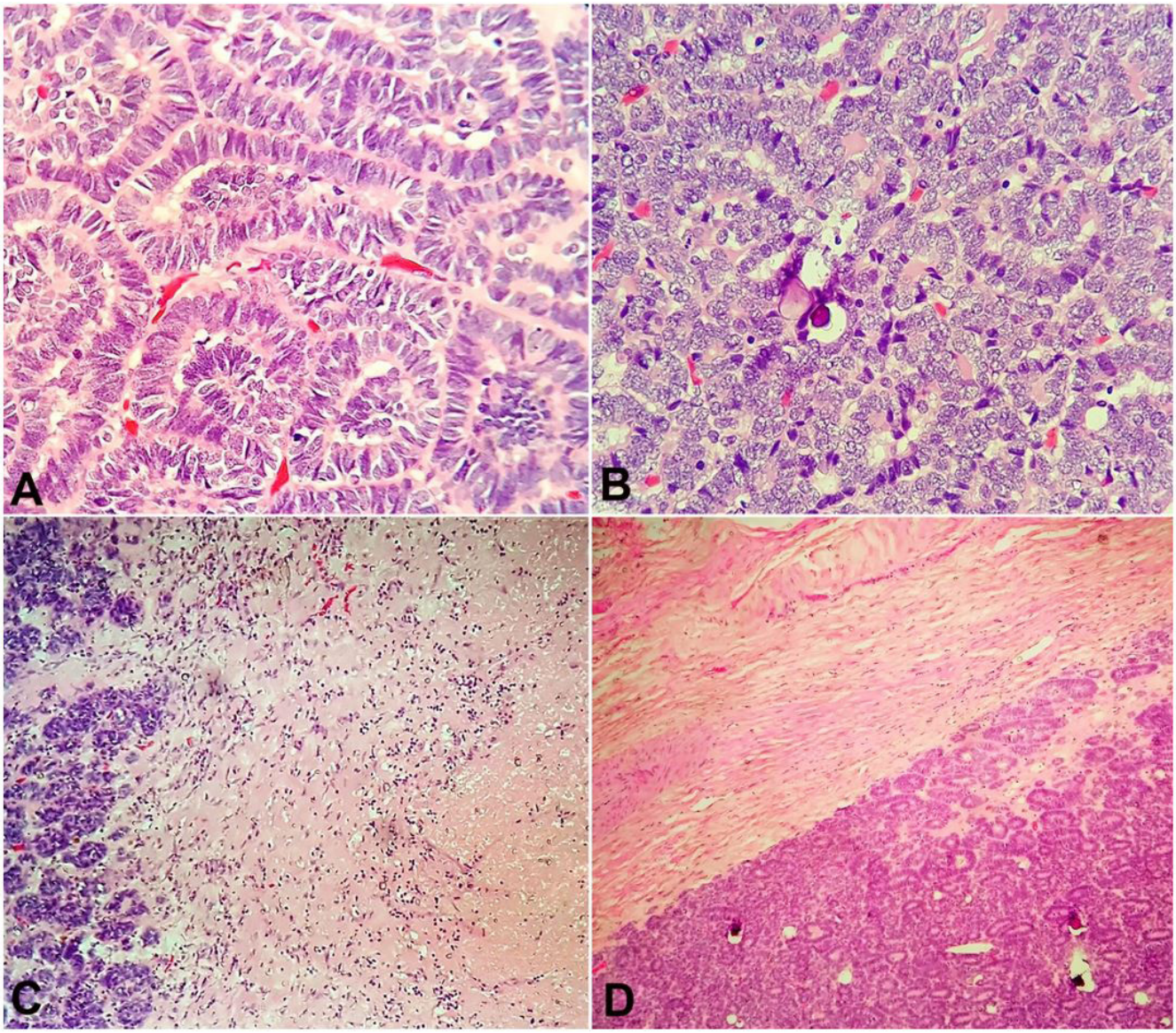

Figure 3. Photomicrographs of the tumor showing **A** and **B –** tightly packed acini and papillary structures with psammoma bodies (H&E, 400X); **C –** area of necrosis (H&E, 100X); **D –** tumor with pushing margin (H&E; 200X).

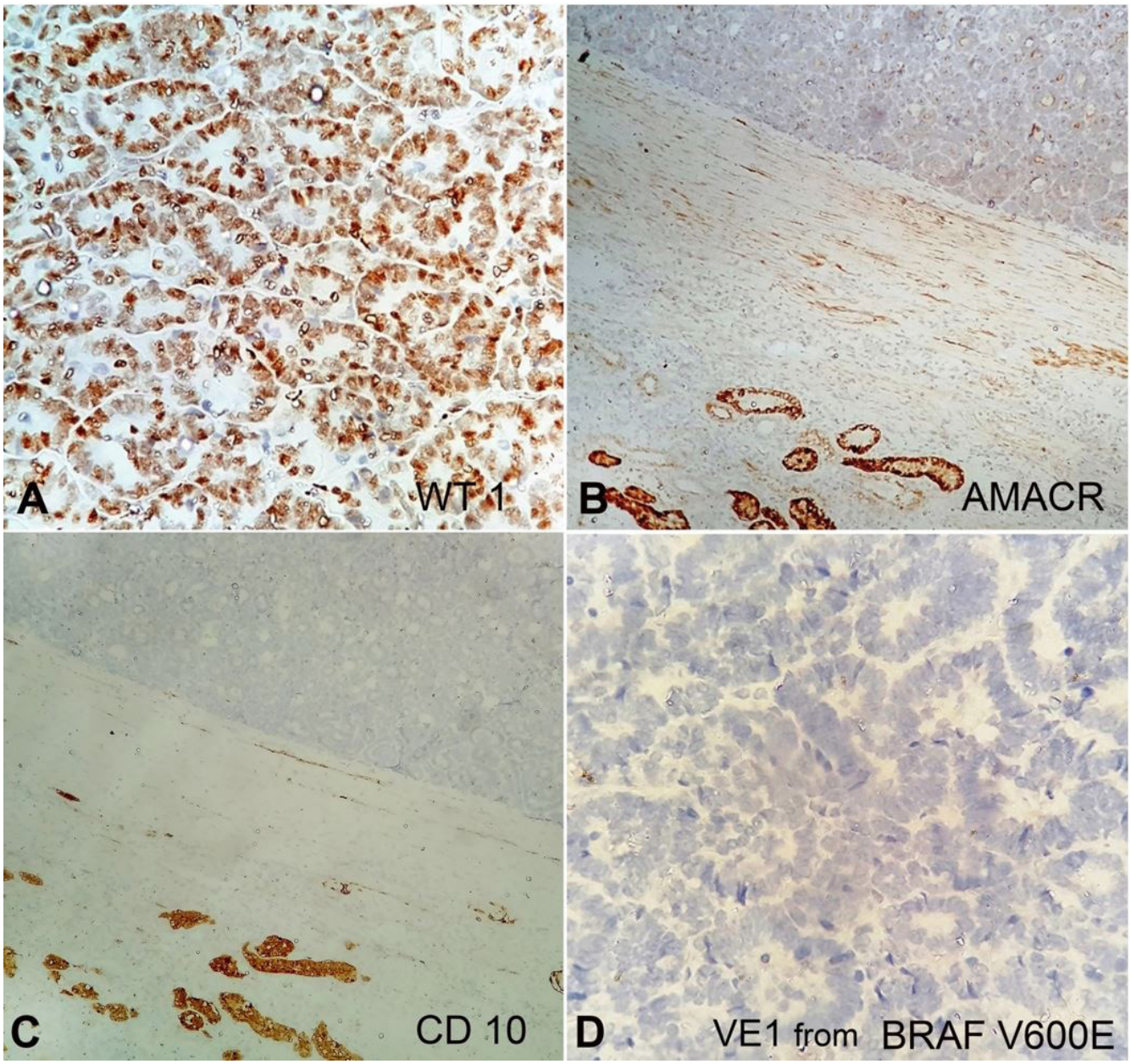

Figure 4. Photomicrographs of the tumor showing in: **A –** positive reaction of the tumor cells for WT1 (400X); **B –** negativity of the tumor cells to AMACR, but positivity in the normal renal tubules (200X); **C –** negativity of the tumor cells to CD10, but positivity in the normal renal tubules (200X); **D –** negative reaction of the tumor cells to VE1 protein (400X).

morphology with epithelial predominant Wilms tumor, while in adults (≥30 years) MA can mimic the papillary renal cell carcinoma morphologically.[7] Clinically, MA is most often asymptomatic and diagnosed incidentally on imaging studies, but may present as an abdominal mass accompanied by flank pain and hematuria.[4] Radiologically, these tumors are well defined, solid mass lesions without enhancement in the posterior aspect of the kidney as viewed in contrast-enhanced CT images.[6] However, it is still difficult to distinguish MA from other entities just by imaging features alone.[3]

Currently, histopathological and immunohistochemical studies are the gold standards to make a definitive diagnosis.[3] On the gross examination, MA is typically well-circumscribed, un-encapsulated, solid masses with the size varying from 0.3-15 cm.[8] The typical feature, noted in our case, is the variegated appearance of the tumor, with solid and cystic areas. Hence, our case was resembling a pRCC that usually present as solid cystic masses in the imaging studies.

Histopathologically, these neoplasms are composed of small, packed uniform and overlapping

epithelial cells with scant cytoplasm, inconspicuous nucleoli, and essentially without mitotic figures. These cells are arranged in varied architectural patterns, including simple tubules, acini, and solid patterns.[1] The tumor cells may proliferate forming glomeruloid bodies with papillary projections. The stroma is usually acellular, edematous, and may show myxoid degeneration.[4] It is different from Wilms tumor as it does not contain the blastemal component and lacks atypia and mitotic figures.[8] In the present case, atypical features and mitoses were absent.

On immunohistochemistry, MA shows diffuse and strong expression for WT-1 and CD57. A low Ki-67 and negative staining for CD56 differentiate this tumor from e-WT. AMACR, CK7, and CD10 are usually not expressed, which excludes the diagnosis of pRCC.[4]

Regarding molecular and genetic studies, the BRAF V600E gene mutation and 2p16 gene changes are the most common findings in the MA.[2] The positive expression of the VE1 protein is correlated with the mutated BRAF gene.[1] Less than 10% of cases of MA do not express this specific gene mutation and are considered as the BRAF-wild type MA, as observed in our case.[2] However, 20% of cases of BRAF-mutated MAs do not express the VE1 protein and may present the mutation if searched with other methods.[1] Caliò et al.[1] discovered that the BRAF-mutated MA was associated with older patients, whereas the BRAF-wild type was detected in younger patients, which is consistent with our findings. With regard to gender, the present case was also in correlation with the findings of Caliò et al.,[1] who demonstrated a strong male predominance in MA that are BRAF-wild type. The presence of a solid architecture along with psammoma bodies and hyalinized stromal background were found to occur more frequently in BRAF-wild type cases.[1] Hence, from the present study, we have derived certain clinicopathologic patterns associated with BRAF-wild type MA. These include younger age group, male predominance, and the presence of a solid component with psammoma bodies.

CONCLUSION

In our present case, we demonstrated a BRAF-wild type MA that presented in a young male patient and exhibits a solid pattern with psammoma bodies on histology. Distinguishing these tumors from its mimickers will help prevent unnecessary extensive treatment as MA treated with complete or partial nephrectomy has an excellent prognosis.

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The authors retain an informed consent signed by the patient authorizing the data publication, and the manuscript is by the Institutional Ethics Committee.

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