Case Report

A RARE CASE OF METANEPHRIC ADENOFIBROMA IN A YOUNG ADULT IN ZARIA: A CASE REPORT AND REVIEW OF LITERATURE

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Abstract

Metanephric neoplasms are rare renal neoplasms identified in both pediatric and adult populations. Metanephric adenofibroma (MAF) is a rare, biphasic, benign tumor containing both stromal and epithelial components and could be potentially mistaken as Wilms' tumor (WT) or Renal cell carcinoma. Patients with MAF have ranged from 13 months to 36 years (median 28 months) with no sex preponderance. Here we report a case of MAF in a young woman. A 25-year-old female was referred on account of 3 years history of recurrent right flank pain, a year history of hypertension and microscopic hematuria. Radiological results showed a fairly circumscribed tumor in the lower pole of the right kidney and serial packed cell volume showing an elevated hematocrit count for her age. A right radical nephrectomy was performed and pathology showed a well circumscribed variegated tumor in the lower pole with histology of a biphasic proliferation of bland acinar structures and myofibroblastic nodules. The blood pressure and packed cell volume reduced to normal levels following surgery. No adjuvant chemotherapy was given. In conclusion, MAF is a very rare renal tumor found in both children and young adults that falls into the larger category of metanephric tumors. The radiologic appearances of MAF are non-diagnostic and impossible to distinguish from other solid renal tumors. Histology is however diagnostic and prognosis is good with resolution of symptoms following radical nephrectomy.

Keywords: Metanephric adenofibroma, polycythemia, Hypertension, Hematuria, Prognosis

INTRODUCTION

Metanephric adenofibromas (MAF) are rare, biphasic, benign tumors containing both stromal and epithelial components in various proportions. [1] MAF was formerly designated nephrogenic adenofibroma, and was first described by Hennigar and Beckwith in 1992.[2,3] This tumor is classified among the metanephric neoplasms, which also include metanephric adenoma (MA) and metanephric stromal tumor (MST). MA is a purely epithelial lesion and MST is a purely stromal lesion, while MAF lies in a spectrum between the MA and MST and could also merge with Wilm's tumor (its malignant counterpart) thus it supports the concept that these are all related lesions. [4] The tumor appears to affect predominantly young patients (13 months to 36 years, median 28 months) with no sex preponderance. [2]

The epithelial component of this tumor presents as tubules and/or delicate papillary structures with very little intervening stroma and commonly show diffuse positivity for AE1/AE3, WT1, CD57, and BRAF V600E while the stromal component commonly shows vimentin and CD34 expression with variable expression of WT1and BRAF V600E. [5] The imaging

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appearance of MAF is nonspecific, with the tumor often resembling Wilms tumor. [4] The management of this tumor is quite different, thus correctly diagnosing MAF may spare the patient toxic adjuvant chemotherapy. Currently, confirming its diagnosis mainly depends on postoperative pathological assessment. In this report, we describe the clinical, radiologic, and pathologic features of MAF occurring in a young adult female.

CASE REPORT

A 25-year-old female presented with a 3-year history of recurring right flank pain with no associated flank swelling, history of gross hematuria, weight loss, and no lower urinary tract symptoms. She was diagnosed hypertensive over a year ago when she was managed for acute bacterial meningitis (which fully resolved); and has been regular on antihypertensives but had experienced poor control.

Physical examination revealed a raised blood pressure (190/130mmHg), with displaced apex beat, but normal heart sounds. Her abdomen was symmetrical with bilateral ballotable kidneys but moderately tender on the right. Ultrasonography revealed a fairly rounded heterogeneous mass at the inferior pole of the right kidney measuring 3.5x4.3cm in dimension with multiple punctate calcifications within it and marked vascularity on color Doppler interrogation; mild hydronephrosis of the renal calyces was also noted with no evidence of invasion of the renal capsule or vessels. A computerized tomographic scan showed an isodense heterogenous soft tissue mass in the mid and lower pole of the right kidney, preserved paranephric region with no abdominal lymphadenopathy. A normal functional left kidney was noted. A diagnosis of a right, Renal cell carcinoma (RCC); Stage T1b Nx Mo was made.

Pre-operative laboratory investigations revealed a raised hematocrit 47.3% (normal, 35%-43%), mild dehydration (mildly elevated urea and sodium with lowered bicarbonates), microscopic hematuria, and urinary casts. Other parameters in the full blood count and electrolytes, the erythrocyte sedimentation rate, liver function tests, random blood glucose, urine culture, hepatitis screen (HBSAg & HCV), HIV screen, and tuberculosis screen were normal or negative.

She was counselled on her diagnosis and treatment and had an open right radical nephrectomy via a mini 12 rib approach following optimization and a written informed consent. Intra-operative findings were that of a tumor at the inferior pole of the right kidney and a grossly normal appearing parenchyma in the middle and superior pole. The liver, peritoneum and retroperitoneum were also grossly normal. The postoperative recovery was satisfactory with a drop in blood pressure to 120/90 mmHg. She was taken off antihypertensives and her blood pressure has remained within the normal range. Follow-up blood pressure and hematocrit were 110/80 mmHg and 35.6%.

Gross pathologic examination of the nephrectomy specimen revealed a right kidney that had a pale multinodular mass close to the lower pole that measured 4 cm x 3 cm x 2 cm with a yellow solid to hemorrhagic cut surface (Fig. 1).

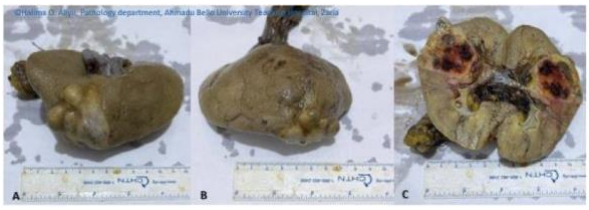

Fig. 1: Metanephric Adenofibroma (Gross). A and B: anteriolateral and posteriolateral views; C: Cut section through the kidney and tumour

The tumor extended from the cortex into a renal papilla. The overlying capsule stripped with mild difficulty showing granular sub capsular surface. There was no extension into the pelvis nor into the renal vessels. Microscopy showed a circumscribed but unencapsulated biphasic tumor composed of nodules of proliferating plump fibroblasts (without collagenization) and delicate branching acini structures lined by bland epithelial cells and containing mucinous material. Mitotic figures were not noted (Fig. 2).

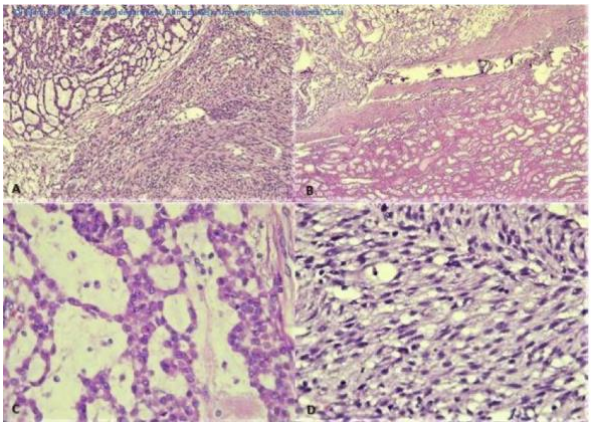

Fig. 2: Metanephric Adenofibroma (H&E). A (x100) Low power view showing both epithelial and mesenchymal components of the tumor; B (x40) Low power view showing the tumor interfacing with remnant renal tissue; C (x400) high power view of the epithelial component; D (x400) High power view of the mesenchymal component.

DISCUSSION

MAF are benign tumors that have been detected incidentally, [4] [6] on investigation of polycythemia and hypertension in the young, [2] mimicking a Wilms tumor, [1] or mimicking a RCC [2] as was the case in this patient. Reports of this tumor are rare with only 25 original articles and case reports detected in the English literature upon a PubMed search as at the time of writing this report. MAF have been reported mostly in children and young adults showing no significant gender preponderance. [3, 5] Together with other metanephric tumors (MA and MST), they have been described as the hyperdifferentiated benign end of Wilms tumor. [3]

In MAF, patients may be asymptomatic but may present with gross hematuria especially when the lesion penetrates the renal collecting system. Microscopic hematuria, polycythemia, and hypertension as was seen in this patient are also seen. These resolved following nephrectomy. Nephrectomy avails the surgeon the opportunity to give a complete treatment as clinical findings and imaging studies cannot differentiate MAF from its malignant differentials. [6] Although total nephrectomy is the treatment of choice especially for large tumors, partial nephrectomy and laparoscopic partial nephrectomy are modalities that have been employed in its management. There is no role for chemotherapy or radiotherapy. [4]

The gross findings here of an unencapsulated variegated solid and cystic tumor is no different from the findings in RCC and Wilm's tumor which are malignant conditions. Histology remains the only means of distinguishing MAF from other similarly presenting tumors postoperatively. Unencapsulated but well circumscribed, it arose centrally, and was composed of a biphasic proliferation of benign epithelial (acinar) structures and nodules of myofibroblasts, with low proliferative indices; other patterns reported include tubules, nests and papillae. This tumor is different from the purely stromal (MST) and purely epithelial (MA) tumors. Although symptoms resolve following excision, long term follow up to monitor renal function is advised. This tumor is not known to recur or metastasize. [3]

In conclusion, MAF is an extremely rare tumor that should be considered in the differential for a non–aggressive appearing solitary renal mass presenting in a young adult who may be asymptomatic. The radiologic appearances of MAF are non-diagnostic. Histology is diagnostic and prognosis is good with resolution of symptoms following radical nephrectomy.

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