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Case Report Open Access

Largest Metanephric Adenoma Kidney with Polycythemia Incidentally Detected in a Blood Donor

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Abstract

Metanephric adenoma of kidney is a rare entity. It presents in young individuals and its association with polycythemia is well described. Differentiation from Wilm's tumor and renal cell carcinoma is challenging and hence definitive diagnosis is made by histopathology with immunohistochemistry. Recognition of this entity is important as it has a more favourable clinical outcome compared with Wilms' tumor and renal cell carcinoma. We report a case of largest metanephric adenoma presenting with polycythemia incidentally detected in young female during blood donation and managed by radical nephrectomy.

Keywords: Metanephric adenoma; Polychythemia; *BRAF*

Introduction

Metanephric tumors of kidney represent a spectrum which contains renal epithelial or stromal cells or both. Metanephric adenoma (MA) is a rare benign neoplasm that has an excellent prognosis. It accounts for only 0.2% of adult renal epithelial neoplasms. The majority of cases occurs in patients 50-60 years of age and is seen predominantly in females by a 2:1 ratio. Clinically pain and palpable mass is most common presenting feature. Polycythemia is associated in 12% of cases which is higher than with any other renal neoplasm. Few cases of metastatic disease have been reported, though most of the cases are benign. The differential diagnosis includes Wilm'stumor, metastatic lung carcinoma, and papillary renal cell carcinoma. Management approach remains surgical resection with definitive diagnosis achieved by histology and immunostaining.

Case Report

A 21-year-old female, dentist my profession was incidentally found to have very high haemoglobin level (19 g/dl) on routine blood donation. On further evaluation, she had raised erythropoietin levels (64.90 IU/L, normal range 3.7–36 IU/L). Rest of the haematology and biochemistry investigation was normal. Haematology consultation was taken. USG abdomen was done which showed large discrete, sharply marginated hypo echoic mass arising from upper pole of right kidney. The size of the mass was 18 x 14 cm. Contrast enhanced CT Abdomen confirmed presence of large hypodense mass (Hounsfield units 5–28) arising from upper and mid pole of right kidney of size 21 x 13 x 16 cm extending superiorly to displace the liver under surface (Figures 1 and 2).

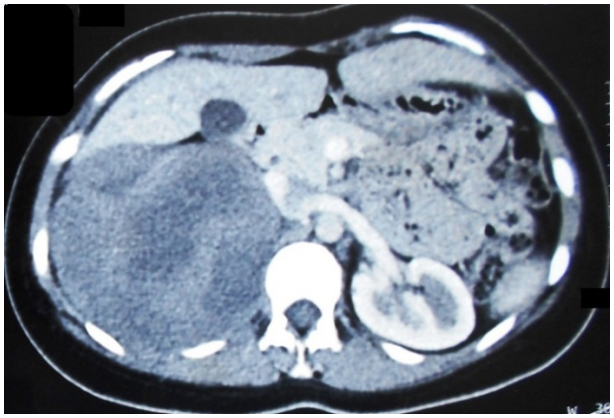

Figure 1: Coronal CT images showing large hypodense mass arising from upper and mid pole of right kidney (HU–24).

The mass showed faint arterial enhancement and was confined in the Gerota's fascia. In view of large renal mass with Polycythemia and CT imaging features, possibility of Wilms tumour, papillary RCC and Metanephric adenoma was considered. USG guided biopsy of mass was done which showed tumor cells arranged in tubular pattern with little intervening stroma. The tumour cells had round to oval perpendicularly oriented with fine nuclear chromatin. No blastemal component was seen. Immunohistochemistry showed tumour positivity for WT-1 with overall features suggestive of metanephric adenoma. Patient underwent open right radical nephrectomy *via* subcostal incision. Intra Postoperative findings were, a well encapsulated renal mass was seen arising from upper pole of right kidney, planes with liver under surface was preserved, there were 2 renal arteries and 1 renal vein, which was ligated separately and cut. Gross examination revealed a well circumscribed mass in the upper

pole of right kidney measuring 14 cm in diameter, greyish white in colour with areas of haemorrhage.

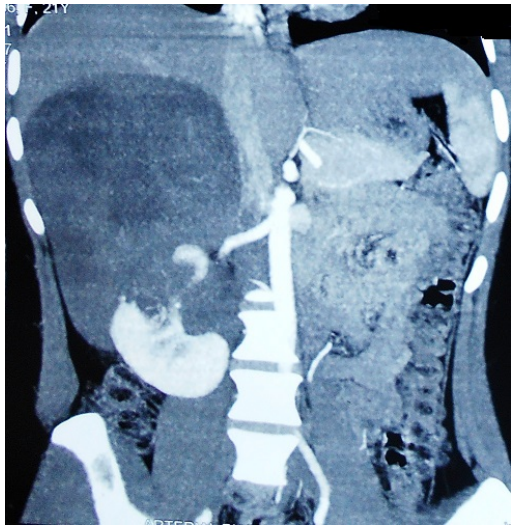

Figure 2: Axial CT images showing large hypodense mass arising from upper and mid pole of right kidney.

The renal cortex was preserved. The adjoining renal parenchyma and hilar vessels appeared grossly unremarkable (Figure 3).

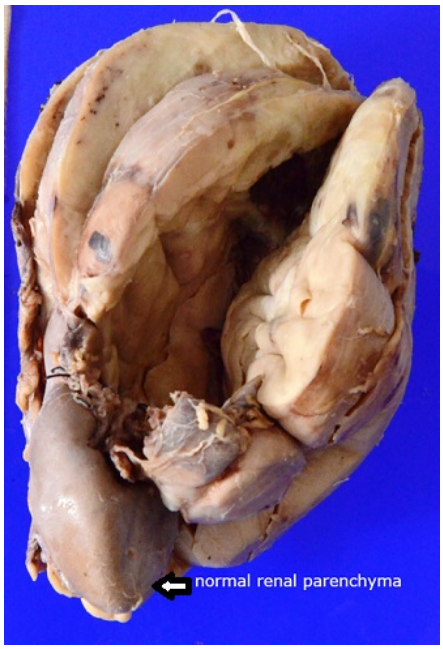

Figure 3: Gross specimen of radical nephrectomy showing bosselated irregular mass with normal lower pole parerchyma.

Microscopically tumour was composed of complex tubular and tubulo-papillary architecture with little intervening stroma. Few areas showed glomeruloid-like bodies. The tumour cells were monomorphic with round to oval nuclei, fine chromatin and minimal cytoplasm. No atypia or mitosis was noted. There was no infiltration into renal sinus, renal capsule and perinephric fat. Lymphovascular and ureteric resection margins were free. On immunohistochemistry, these tumor cells showed nuclear positivity for CD-57 and WT-1, and negative for AMACR and CK-7 (Figure 4).

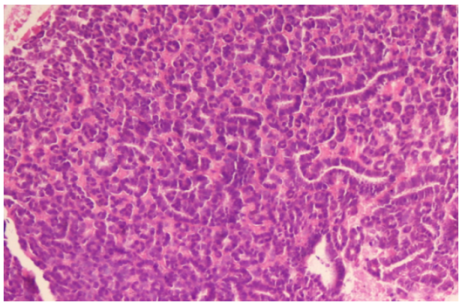

Figure 4: Photo micrograph shows a tumour composed of complex tubular and tubulo-papillary architecture with little intervening stroma.

Further, molecular testing for *BRAF* gene mutation using Sanger's sequencing technique was done which showed positive result for *BRAF*-V600E mutation, suggesting as metanephric adenoma (Figure 5). Post op period was uneventful. Her haemoglobin was 13.4 g/dl and erythropoietin levels came to normal range. Patient was discharged on day 5 after surgery. She is doing well on regular follow up for the last 18 months without any evidence of recurrence.

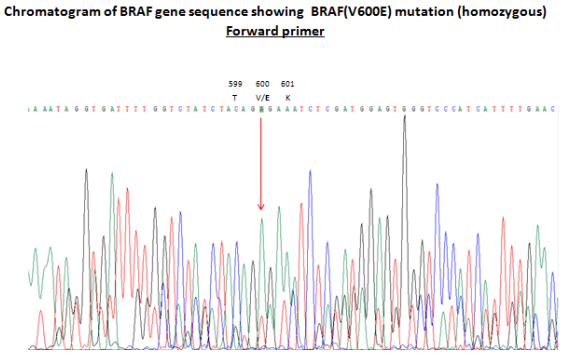

Figure 5: C Chromatogram of *BRAF* gene Sequence showing *BRAF* (V600E) mutation (homozygous)- Forward Primer.

Discussion

Metanephric Adenoma (MA) is a rare neoplasm, characterised by presence of spindle cells along with epithelial cells [1]. It has a peak age of occurrence in the fifth or sixth decade of life with female preponderance. Clinically it may be asymptomatic, or present with symptoms like abdominal pain, abdominal mass, hematuria, dysuria, fever, or hypertension. MA has the highest incidence of polycythaemia (12%) among all renal lesions [2]. Polycythemia may represent a paraneoplastic syndrome of this rare entitiy. In the large series of 50 cases of Metanephric Adenoma reported by Davis et al. there was a distinct female preponderance, with an approximate female: male ratio of 2:1. The most common presenting symptoms reported was

abdominal pain and hematuria and mean tumour size was 5.5 cm, with a range of 0.3-15.0 cm [3]. Currently, neither ultrasound nor CT scans can reveal distinct features of MA. Ultrasound scans show both hyper-echoic and hypo-echoic regions, while CT scans show a non-distinct mass with low attenuation on contrast studies [4]. In our case, renal mass was hypo echoic and well marginated on USG and CECT abdomen showed hypodense mass with few areas of enhancement suggesting a possible differential diagnosis of Renal cell cancer. Most MA reported in literature has mean size of 4–8 cm. Most of the cases are unilateral and unifocal. The index case was unique being multifocal and largest size yet reported. MA tumours appear tan or gray in colour with and may be soft or firm. Foci of necrosis and hemorrhage are common. Calcifications are uncommon, and only occur in approximately 20% of cases [5]. Histologically, MA is composed of tightly packed uniform small epithelial cells with small regular nuclei that form a tubular or acinar pattern, a high nuclei-to-cytoplasm ratio, and no mitotic figures. The histological resemblance of renal MA are Papillary renal cell carcinoma (PRCC) and epithelial Wilm'stumor [6,7]. However wilms tumour is seen more common in pediatric age group. Immunostaining is sensitive tool in differentiating MA from the other counter parts. While MA shows diffuse positive staining for CD57 and WT1 and weakly for CK7 and EMA, PRCC shows strong positivity for CK7 and EMA immunoreactins and wilms tumour is characterized by epithelial, stromal, and blastemal components. *BRAF* encodes a serine/threonine specific protein kinase upstream of the MAPK/ERK signalling pathway. The V600E mutation of *BRAF* is present in approximately 90% of Metanephric Adenoma serving as a potential valuable diagnostic tool in differential diagnosis [8]. We further did molecular testing of V600E mutation of *BRAF* using Sanger sequencing analysis and it was positive for the tumour cells suggesting it a MA.

Although MA is usually benign, a few cases of metastatic disease have been reported [9] MA is a benign lesion; however it should always be resected in order to confirm the diagnosis and rule out PRCC and other rare malignant counter parts of renal cancer. In our case, the patient was young and healthy and she had developed polycythemia which made the resection mandatory. Immunochemistry and molecular testing is essential to diagnose this rare benign renal tumour.

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