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Case Report

Pathology Section

Diagnostic Conundrum in a Case of Solid Papillary Renal Cell Carcinoma

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ABSTRACT

Papillary Renal Cell Carcinoma (RCC) is the second most common carcinoma arising from the renal tubular epithelium. It comprises 10-15% of cases in surgical series. The solid variant of papillary RCC includes only those RCCs that lack true papillae but contain the characteristic immunohistochemical and genetic features of papillary RCC. Here, we present the clinical, histopathologic and immunohistochemical findings of a rare case report of solid variant of papillary RCC. A 45 years old female presented with left abdominal mass. Histopathological sections showed tumour cells arranged in tubules, trabeculae and cords with occasional interspersed long branching and angulated channels with pointed ends. Even extensive sampling of the tumour failed to reveal any papille. The differential diagnosis included solid papillary RCC (s-PRCC), Metanephric Adenoma (MA) and Collecting Duct Carcinoma (CDC). Histologically, these tumours may show overlapping features. The use of IHC markers panel comprising of CK7, EMA, Vimentin, AMACAR and WT1 can help to reach at a diagnosis.

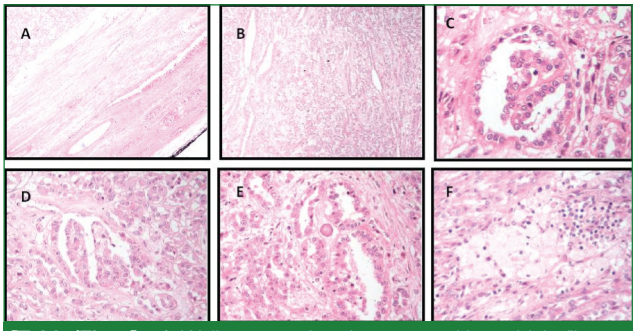
Keywords: Collecting duct carcinoma, Metanephric adenoma, Solid renal cell carcinoma

CASE REPORT

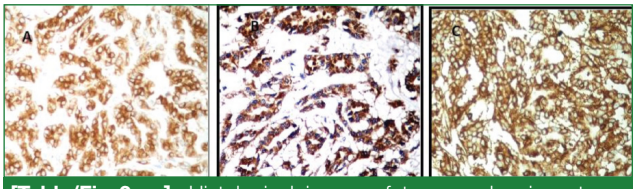
A 45-year-old female presented with left abdominal mass and pain for one year. Her medical history and physical examination were unremarkable except for a bimanually palpable, ballotable, firm mass measuring 9X8 cm in left lumbar region, extending into umbilical region, part of left iliac fossa and hypogastrium. Possibility of renal cell carcinoma was considered. Computed tomography scan demonstrated a large encapsulated well defined heterogeneously enhancing mass lesion measuring 8.7 x 8.3 x 7.6 cm arising from lower pole of left kidney. The right kidney and rest of the abdomen was unremarkable. After taking consent from the patient a left sided radical nephrectomy was performed.

Grossly, the kidney measured 15 x 9 x 8 cm. The cut surface showed a homogenous, tan brown well circumscribed tumour at the lower pole of the kidney measuring 9 x 8 x 6 cm. The tumour was confined within the renal capsule.

Microscopically, the tumour was separated from the surrounding renal parenchyma by a thick fibrous pseudocapsule [Table/Fig-1a]. The tumour was composed of predominantly of tubular pattern with few areas showing cells arranged in trabeculae and cords. Occasional interspersed long branching and angulated channels with pointed ends were also noticed [Table/Fig-1b]. The Cells were cuboidal with moderate amount of eosinophilic cytoplasm, round nucleus with mild anisonucleosis and fine granular chromatin. Few cells showed prominent nucleoli. Even extensive sampling of the tumour failed to reveal any papillae with fibrovascular core



[Table/Fig-1]: a) Well encapsulated tumour with a thick fibrous capsule (H&E 100X); b) Tumour cells were arranged in tubules with occasional interspersed Long branching and angulated channels with pointed ends (H&E 100X); c) Few glomeruloid structures (H&E 400X); d) Pseudopapillae (H&E 400X); e) Occasional psammoma bodies (H&E 400X); f) Collections of foam cells (H&E 400X).



[Table/Fig-2a-c]: Histological image of tumour showing strong positivity to CK 7 (400X).

or areas of necrosis. However, few glomeruloid structures, pseudopapillae and occasional psammoma bodies were noticed along with foci of microcalcification and collection of foam cells [Table/Fig-1c-g]. By immunohistochemistry, the

tumour was strongly positive for CK-7, vimentin, EMA and AMACAR but negative for S-100 and WT1 [Table/Fig-2a-c]. Based on morphological findings and immunohistochemistry a final diagnosis of solid papillary renal cell carcinoma was rendered. The post-operative course was uneventful. Last follow-up, 18 months after excision, the patient was well and without any evidence of metastasis.

DISCUSSION

Papillary RCC is the second most common carcinoma arising from the renal tubular epithelium and comprises 10-15% of cases in surgical series [1]. To be called as a papillary RCC, at least 75% of the tumour should be composed of papillary or tubulopapillary histology with fibrovascular cores [2]. Several subtypes of papillary RCC had been described [1]. In 1997, Renshaw AA et al., in their series of 6 cases defined the solid variant of papillary RCC as those RCCs that lack true papillae but contain the characteristic immunohistochemical and genetic features of papillary RCC [3]. Although, first described in 1997, to best of our knowledge, only few cases have been described so far [4-7].

Histopathological sections from papillary RCCs show papillary or tubulopapillary structures lined by small, cuboidal cells with basophilic cytoplasm or by larger cells with eosinophilic cytoplasm [1]. Collections of foamy macrophages within the papillary stalks, iron deposition, and psammoma bodies are a frequent feature of these tumours. Although, papillary RCC was originally defined histologically, these tumours also have characteristic immunohistochemical features. Papillary RCC is typically immunoreactive for cytokeratin 7, cytokeratin 8/18, CD10, epithelial membrane antigen vimentin and a-Methylacyl co-enzyme A racemase [8]. Papillary RCC often show foci of solid areas admixed with tubulopapillary areas. In this report, we describe a tumour with a solid and tubular growth pattern completely lacking true papillae or spindle cell areas. The tumour retained the immunohistochemical findings of papillary renal cell carcinoma and showed immunoreactivity for CK-7, vimentin, EMA and AMACAR but was negative for WT 1 and S100.

The main differential diagnosis of s-PRCC includes Metanephric Adenoma (MA) and Collecting Duct Carcinoma (CDC).

Although, MA and s-PRCC share some morphological features, some architectural and cytological characteristics may be useful in their distinction [7] [Table/Fig-3]. MA cells are uniform, small, with high nuclear: cytoplasmic ratio, and show bland nuclei with delicate chromatin and absent or inconspicuous nucleoli. In contrast, the cells in s-PRCC are less uniform, with a variable nuclear: cytoplasmic ratio and occasional larger cells containing larger amounts of cytoplasm. The nuclei shows open or vesicular chromatin and nucleoli are present. Mitosis is very rare or absent in MA.

In particularly challenging cases, the use of IHC markers may be an important aid in reaching an accurate diagnosis. MA show strong positivity for WT1 and are negative for CK7, EMA, vimentin and AMACAR. The reverse holds true for solid papillary renal cell carcinoma. Histologically, CDC often has a mixed papillary and infiltrative tubular architecture. The infiltrative component is associated with marked stromal desmoplasia. Foci of dysplasia, or carcinoma in situ, can be found in the adjacent collecting ducts in some cases. The tumours are of high nuclear grade, corresponding to Fuhrman grade 3 or 4 [8]. On immunohistochemistry, CDC is strongly positive for Ulex European agglutinin 1 lectin. CDC is also positive for Peanut Agglutinin (PNA), vimentin, lysozyme, distal tubular marker EMA, and high molecular weight cytokeratin, and negative for proximal tubular markers (RCC marker and CD10) [9].

Features	MA	s-PRCC	Present Case
Age (in years)	35-55	50-58	45
M:F	1:1.3	5:1	F
Size	0.2-15 cm	1-12 cm	9 cm
Focality	Unifocal/ Multifocal	Unifocal/ Multifocal	Unifocal
Capsule	Thin discontinuous capsule; No infiltration	Thick fibrous pseudocapsule; may show infiltration	Thick fibrous capsule No infiltration
Tubules ++	++	++	++
Long Branching Channels with Angulated Ends	++	-	Occasional
Pseudopapillae	+	+	+
Glomeruloid Bodies	+	+	+
Psammoma Bodies	+	+	+
Dystrophic Calcification	+	+	+
Foam Cells	-	+	+
Cell	Uniform small cells blan chromatin	Less uniform Open/vesicular chromatin, nucleoli	morphology + Mild pleomorphism, prominent nucleoli In few cells
Mitosis	Rare 0-1/10 HPF	1-2/10 HPF	0-1/10 HPF

[Table/Fig-3]: Key features of Metanephric Adenoma (MA) and solid Papillary Renal Cell Carcinoma (s-PRCC).

CONCLUSION

In this case of RCC, tumour cells were arranged in tubules and focal solid areas. The tumour lacked any true papillae. The tumour showed focal glomeruloid structures. The histopathological findings made it difficult to distinguish it from metanephric adenomas. The immunohistochemical findings showing positivity for CK7, EMA, Vimentin and AMACAR and negativity for WT1 helped to reach the diagnosis of solid papillary RCC. Despite the lack of true papillae, tumour is

best classified as a solid variant of papillary RCC. This case is one of the few cases of s-PRCC reported in the literature.

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