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Case Report

A case report of mucinous tubular and spindle cell carcinoma of the kidney

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Abstract: Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare type of kidney tumor that has only recently been described, with less than eighty cases in the literature. This was only recognized as a specific entity in the World Health Organization 2004 classification of Renal Cell Carcinoma (RCC). MTSCCs are polymorphic renal neoplasms characterized by small, elongated tubules lined by cuboidal cells with cords of spindled cells separated by pale mucinous stroma. We report the case of a 57 year old lady who had an incidental finding of a mass in her right kidney. The radiological features were consistent with a RCC and following a multidisciplinary team discussion she underwent a laparoscopic radical nephrectomy. Macroscopic examination revealed a well circumscribed 6.5 × 6 × 6.5 cm right lower pole mass. Histologically it was composed of elongated tubules, small tubules and papillary structures with a necrotic centre. The cells demonstrated cuboidal and spindle cell morphology. Histological grade was Fuhrman grade 2. The majority of MTSCCs are indolent, and there are only two reports of distant metastases which responded favorably to adjuvant sunitinib. To date there is no international consensus on long term surveillance of these patients. Due of the favorable prognosis with this type of tumor, MTSCC must be differentiated from papillary renal cell carcinoma to avoid administration of excessive adjuvant treatment to patients.

Keywords: MTSCC, kidney cancer, papillary RCC with sarcomatoid differentiation, metanephric adenoma

Introduction

Mucinous tubular and spindle cell carcinoma (MTSCC) is an exceedingly rare form of renal cell carcinoma [1]. Although only first recognised as a specific entity by the World Health Organisation in 2004, it is regarded as a low grade renal epithelial neoplasm, due to the negligible mortality rates associated with this malignancy [2]. MTSCC has a characteristic histological appearance which has been well documented. It comprises small elongated tubules, lined by cuboidal cell with cords of spindled cells separated by pale mucinous stroma.

However it must be noted that MTSCC shares a number of histological features with other more virulent forms of renal cell carcinoma and thus precise diagnosis is paramount particularly with regard to the necessity for adjuvant/neoadjuvant therapy and subsequent prognosis.

Case report

This case centers on a 57 year old previously healthy lady, who was referred to the Urology Service with an incidental ultrasound finding of a large mass in her right kidney. Past medical and surgical history was of no significance. Initial hematological and biochemical investigations were within normal range. Subsequent CT Thorax Abdomen Pelvis demonstrated a large (5.8 cm × 5.7 cm × 5.5 cm) peripherally well defined, solid, heterogeneously enhancing mass emanating from the lower pole of the right kidney (**Figure 1**). There was no convincing evidence for metastatic disease.

The case was discussed at a multi-disciplinary team meeting, a preliminary diagnosis of renal cell carcinoma (RCC) was made and following discussion with the patient, radical nephrectomy was scheduled. The post operative period was uneventful and the patient was discharged

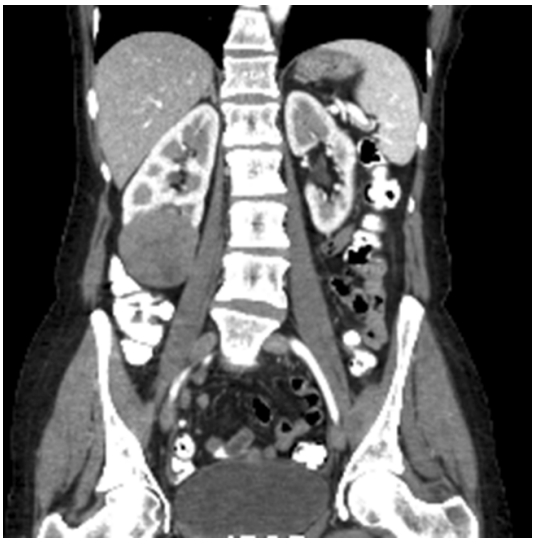

Figure 1. CT Abdomen revealing a ~6 cm right lower pole renal mass.

home well. Subsequent gross examination of the resected specimen revealed a large mass with a cream cut face, which was centrally necrotic. It extended into, but did not invade the renal pelvis.

Histological examination demonstrated a tumor composed of elongated tubules, small tubules and papillary structures (**Figure 2**). The cells showed cuboidal and spindle cell morphology without significant nuclear pleomorphism. Only scattered mitotic figures were seen. No significant mucinous component was present. The appearances were of a mucinous tubular and spindle cell carcinoma, mucin poor variant. Prognosis was considered favorable and MDT consensus was for follow up ultrasound after six months moving to annual review thereafter.

Discussion

MTSCC is a rare entity. First described in 1997 it was first recognised as a specific tumor type by the World Health Organisation in 2004 [2]. Its significance lies in the need for accurate diagnosis. MTSCC tends to occur in adults, with case series reporting mean ages of 52-54 years [3, 4]. The reported ratio of males to females is 1:3. Presenting symptoms include flank pain and/or hematuria. Visible hematuria as a presenting complaint is thought to be due to the fact that the tumor often arises from the renal medulla [5]. However, as is the trend in renal masses, the diagnosis is made following an incidental finding on advanced imaging.

MTSCC has a characteristic appearance. Grossly the tumor is often quite large, and may have a necrotic component [6]. Histologically, it exhibits a mixed pattern of tubules and a surrounding spindle cell proliferation within a myxoid structure. Importantly mitotic figures are infrequently encountered in the histological examination of MTSCC specimens. In general the tumors are confined within the renal parenchyma (pT1 or pT2), with few reports of metastatic disease [6]. The majority of reported cases of nodal or metastatic disease involved tumors with a sarcomatoid component or had a higher nuclear grade [7, 8]. Treatment with adjuvant sunitinib and separately nivolumab plus ipilimumab, induced a favorable response both radiologically and symptomatically in reported cases of distant metastases in patients with MTSCC [9, 10].

A number of histological types are included in the differential diagnosis of an RCC. Distinction between various subtypes is paramount. Accurate diagnosis dictates both prognosis and follow up. A histological mimic of MTSCC is papillary RCC with sarcomatoid differentiation. RCC with sarcomatoid transformation is reported to have poor outcomes, largely owing to the advanced or late-stage of disease at the time of diagnosis. One and five year survival rates of 59% and 22% respectively, have been reported [11]. Fluorescence in situ hybridization (FISH) is crucial to the differentiation of these two tumor types. Full-length gains in chromosomes 7, 12, 16, 17, and 20 are found in papillary RCC, while MTSCC has been demonstrated to have gains in chromosomes 2, 4, 5, 7, 9, 10, 12, 16, 17, 18, 19, 20, 22, and others include losses of chromosomes 1, 4, 6, 8, 9, 13, 14, 15, 22 and X [6, 7]. Other differential considerations include metanephric adenoma, sarcomatoid renal cell carcinomas and collecting duct carcinomas. Differentiating factors include a lack of extracellular mucin in type 1 and 2 papillary RCC, less cytoplasm in the case of metanephric adenoma, increased mitotic activity as found in sarcomatoid renal cell carcinomas and particular cytonuclear atypia shown in collecting duct carcinomas [12].

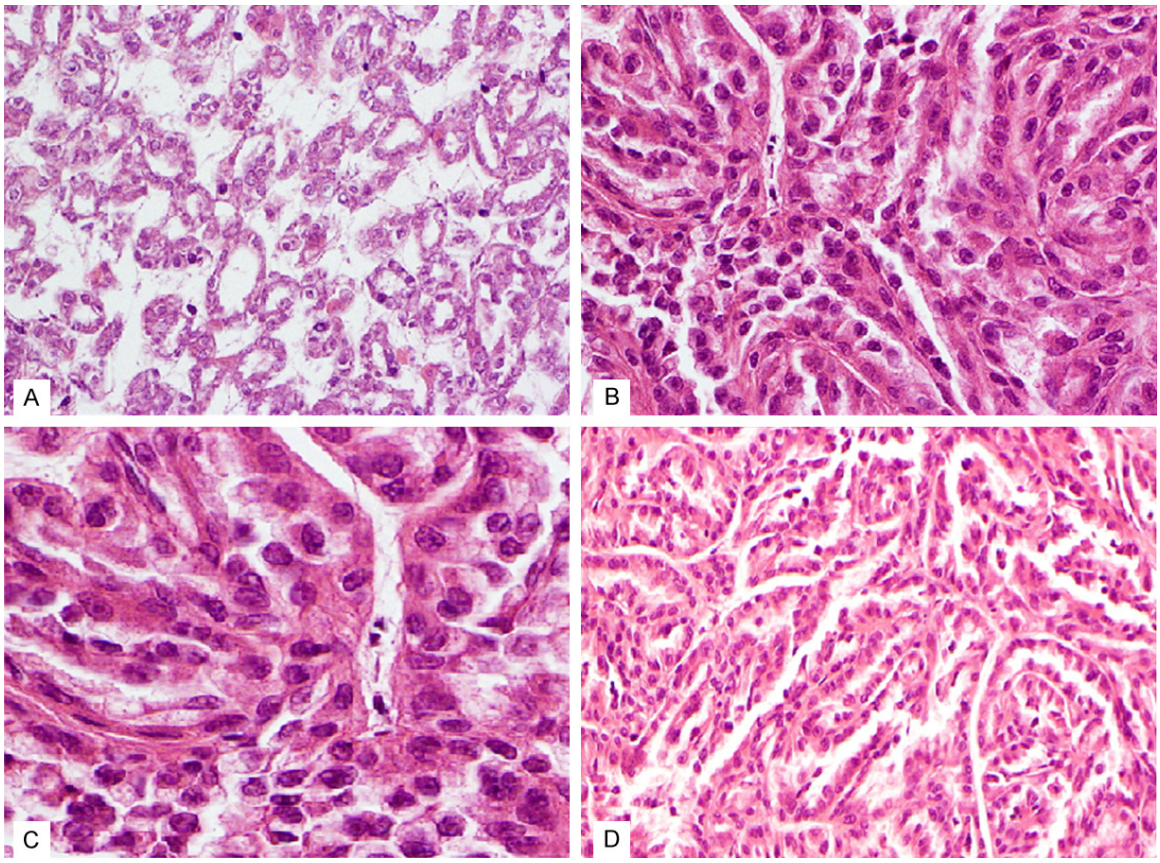

Figure 2. Histological examination demonstrated a tumour composed of elongated tubules, small tubules and papillary structures. The cells showed cuboidal and spindle cell morphology without significant nuclear pleomorphism.

Conclusion

MTSCC is rare, and only relatively recently described, and thus no international consensus exists with regard to patient outcomes, or optimal follow up for this diagnosis. Although MTSCC does display a unique histological profile, there are RCC variants which resemble MTSCC and have a much less favorable outcome. Such entities must be accurately excluded when diagnosing MTSCC. Judicious use of immunohistochemical staining and if needed FISH is recommended in order to manage such cases appropriately. An exact diagnosis is critical to ensure optimum management with regard to prognosis, adjuvant therapy and surveillance.

Disclosure of conflict of interest

None.

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