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Chromophobe Renal Cell Carcinoma with Osteosarcomatous Differentiation: A Rare Case Report

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Abstract: Renal cell carcinoma sometimes exhibit sarcomatoid transformation. Sarcomatoid renal cell carcinoma (SRCC) is an uncommon but not rare neoplasm consisting of a typical renal cell carcinoma (RCC) associated intimately with a sarcomatoid component. It has been reported that sarcomatoid renal cell carcinomas constitute approximately 1.0% to 4.8% of the total number of renal parenchymal tumors. SRCC has an extremely poor prognosis Sarcomatoid transformation has been noted in virtually all types of RCC. However, chromophobe renal cell carcinoma (CRCC) is the most frequent renal cell carcinoma with a sarcomatoid component. Very few cases are reported in the literature, we report this additional with review of literature. A 50 year old woman referred a one month history of constant left flank pain. A left radical nepherectomy was performed. Gross pathological examination showed a tumor located in the upper part of the kidney with variegated appearance. Histologically, the tumor was composed of two intermixed distinct morphologic components; a chromophobe renal cell carcinoma and a high grade spindle cell sarcoma with osteosarcomatous areas. Our case represents sarcomatoid chromophobe renal cell carcinoma. This unusal variant has the potential to behave aggressively and to metastasize.
Keywords: Chromophobecell, Histochemistry, Osteosarcomatous, Renal cell carcinoma, Sarcomatoid differentiation

INTRODUCTION

Chromophobe renal cell carcinoma (CRCC) was first described by Thoenes *et al*. in 1985 [1]. CRCC is a distinct subtype of renal cell carcinoma (RCC) which is a rare variant, with distinct histochemical, ultrastructural, and gentic characteristics. They are relatively uncommon accounting for 4-5% of renal neoplasms. The incidence of sarcomatoid transformation in chromophobe type is about 9 % [2, 3]. The finding of heterologous elements in the sarcomatoid component of CRCC is an extraordinary event, which has been reported in only 3 previous cases. CRCC is characterized by presence of polygonal cells in solid, alveolar sheets with well defined borders, clear to eosinophilic cytoplasm and perinuclear clearing giving plant cell like appearance [4]. Sarcomatoid transformation in this tumor, although rare, has been well documented in the literature and, as in other types of RCC, carries an ominous prognosis for the patient [5]. Here, we present the fourth such case, occurring in the left kidney of a previously healthy 50-year-old woman diagnosed as chromophobe renal cell carcinoma with osteosarcomatous differentiation. The histochemical and immunohistochemical features are presented with review of literature.

CASE REPORT

A 50 Year old woman presented with history of left loin pain since one month. She had a non-contributory medical history. Her physical examination was unremarkable. Lab investigations of renal function test were normal. Ultrasonography of abdomen reported dilated pelvi-calceyal system on left side with echogenic foci noted with multiple calcifications suggestive of infective etiology. An abdominal computerized tomography revealed a non homogenous mass, which measured 9.8cm x 7.8cm, involving the upper pole of the left kidney. The working diagnosis was renal cell carcinoma. She underwent a left radical nepherectomy.

Pathological Findings

Macroscopic Features

A radical nephrectomy specimen consisted of left kidney surrounded by perinephric adipose tissue and Gerotafascia (550 g), with the kidney alone measuring 15.0 x 11 x 8.0 cm and ureter measuring 1cm, renal capsule was breached in areas (Fig. 1). On the cut surface the tumor showed a large circumscribed mass in the upper pole of the kidney and measured

9.8cm x 7.8cm with a variegated appearance, with one area yellowish-white in colour, and the other area showed haemorrhage, cystic change, necrosis and tan in colour. The tumor partly invaded the surrounding adipose tissue. The renal sinus, vascular and ureteral margins were free of tumor (Fig. 2).

Microscopic Features

Histologically, the tumor was predominately composed of two intermixed distinct morphologic components, chromophobe renal cell carcinoma and sarcomatoid neoplasm, with CRCC as the carcinomatous elements and high grade pleomorphic sarcoma as the sarcomatoid component (Fig. 3). The latter displayed areas of osteosarcoma (Fig. 4). The carcinomatous component shows polygonal cells,which were large size with prominent cell membranes and abundant pale pink cytoplasm, with flocculent appearance.

Characteristic perinuclear halos were also present giving the cell a koilocytic appearance. The nuclei were centrally located and had irregular outlines (raisinoid) with different degrees of hyperchromasia. Areas of sarcomatoid and osteiod component were present.

The sarcomatoid cells were Spindle shaped cells arranged in diffuse sheets, fascicles, and storiform like pattern. Bizarre tumor giant cells, many typical and atypical mitotic figures and multinucleation were also seen.

Histochemistry and Immunohistochemistry

The epithelial component was diffusely positive for Hale colloidal iron stain (Fig. 5). Immunohistichemical studies, chromophobe RCC demonstrate pancytokeratin, epithelial membrane antigen positivity [R]. It is also reported that these tumours are positive for CD 117 [c-kit]. The sarcomatoid component demonstrated vimentin.

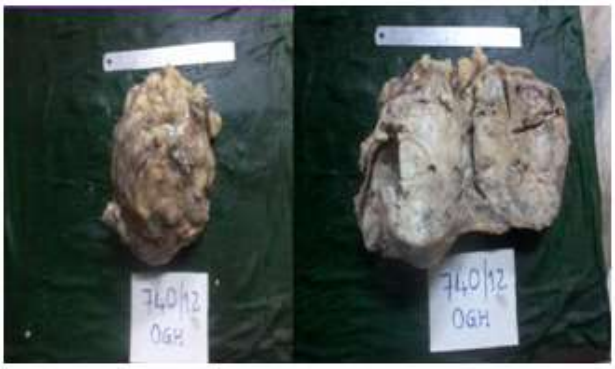

Fig. 1: Macroscopic finding showing surface nodularity and cut surface of tumour with variegated appearance, one area yellowish white in colour, and other area showing with haemorrhage, cystic change and tan in colour

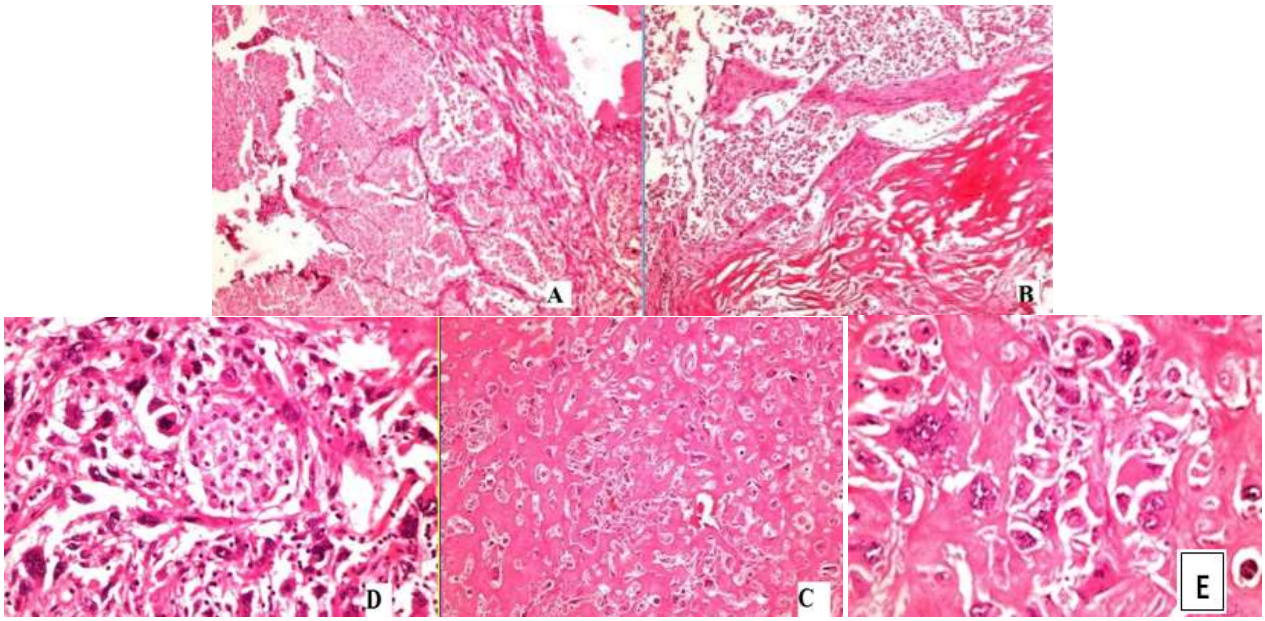

Fig. 2: 10 X A, B, and C, 40X H & E D and E: Low and high magnifications shows tumor with biphasic elements of sarcomatoid CRCC (hematoxylin-eosin), magnifications

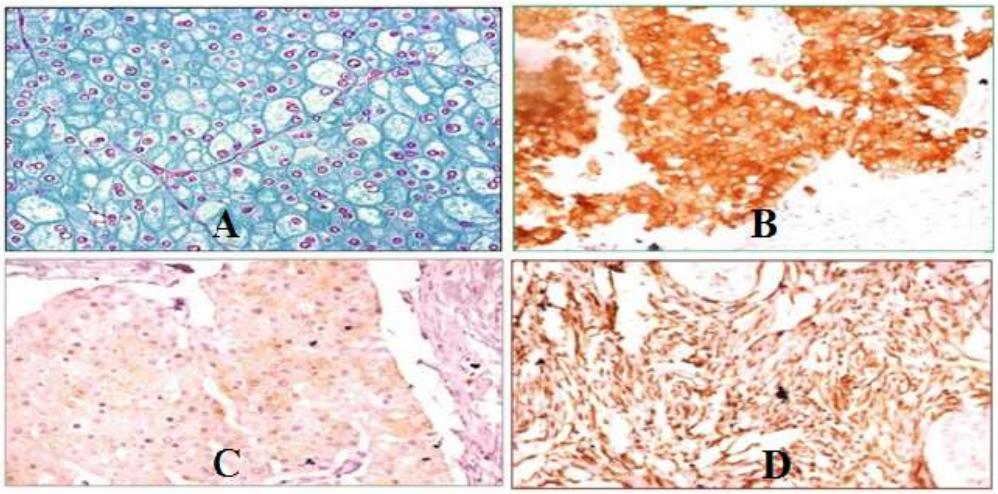

Fig. 3: A: Hales Colloidal Iron-positive, B: EMA-Positive, C: CD 117-Positive, D: Vimentin-Positive

The epithelial component was diffusely positive for Hale's colloidal iron stain. Immunohistichemical studies, chromophobe RCC demonstrate pancytokeratin, epithelial membrane antigen positivity (E29). It is also reported that these tumours are focal positive for CD 117 [c-kit](Polyclonal rabbit). The sarcomatoid component demonstrated vimentin (V9).

DISCUSSION

CRCC is generally diagnosed in the sixth decade of life. It can also present in younger age group of 30-40yrs [5]. Its Incidence is the same in men and in women and 86% usually diagnosed in Stage 1 and Stage 2. It arises from the collecting tubules of kidney. Renal vein invasion presents alone in fewer than 5% of all RCC cases. Metastases are found in 6% of patients and the most frequent sites are the liver (39%) and lung (36%) [3]. Sarcomatoid differentiation is found in 1.9% of CRCC.

In recent classifications, tumors composed of carcinoma and sarcomatous elements are categorized into histological subtypes of the primary carcinoma components. Tumors composed exclusively of sarcomatoid or spindle elements without a recognizable epithelial component are categorized as unclassified RCCs [6].

Histochemical studies shows epithelial component with diffuse positivity for Hale's colloidal iron stain (Fig. 5). Immunohistochemical studies of chromophobe RCC demonstrate pancytokeratin, epithelial membrane antigen positivity. It is also reported that these tumours are positive for CD 117 (c-kit) [6, 7]. The sarcomatoid component demonstrated vimentin.

If these tumors are subjected to ultrastructural and chromosomal analysis, some unclassified RCCs may be able to be categorized into definite histological subtypes. The genetic events responsible for sarcomatoid transformation of RCCs have not been elucidated. Further studies are therefore needed to identify the key gene [8, 9].

Cytogenetic studies of chrromophobe RCC show characteristically extensive chromosomal losses (monosomy) of chromosomes 1,2,6,7,13.17,21, leading to hypodiploidy DNA index. These multiple chromosomal gains may play a role in sarcomatoid transformation of chromophobe RCC [10].

CONCLUSION

In conclusion, considering the prognostic significance of sarcomatoid elements and their proportion in renal cell carcinomas, we emphasize the importance of extensive sampling of tumor mass and search for sarcomatoid elements for proper diagnosis of these highly malignant tumours. It is important to recognize CRCC as a distinct subtype of renal neoplasm; so much as the diagnosis implies a favorable prognosis to the patient. However, if sarcomatoid transformation develops, the patient's prognosis is very poor. Hence awareness of this entity helps in definitive diagnosis and management.

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