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Case Report

The Chromophobe Renal Cell Carcinoma with Sarcomatoid Differentiation

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ABSTRACT

We present a rare case of a 52-year-old woman with a chromofobe renal cell carcinoma that had sarcomatoid components. The tumor, 70x60mm in size, was incidentally found by abdominal ultrasonography. An abdominal computed tomography demonstrated a well-demarcated solid tumor arising from the upper pole of the right kidney. Histologically, the tumor was composed of two intermixed distinct morphologic components: a chromophobe renal cell carcinoma and a high-grade spindle cell sarcoma. There was a sarcomatoid area composed of highly atypical spindle cells intermingled with rhabdoid cells. A unique histological finding of this tumor was the presence of calcification and psammoma body formation throughout the tumor. We think that this case supports the existence of a tumor progression pathway from chromofob to sarcomatoid renal cell carcinoma. A literature review on sarcomatoid chromofobe renal cell carcinoma with calcification was performed

Key words: *Chromophobe renal cell carcinoma, sarcomatoid renal carcinoma, calcification*

Sarkomatoid Komponenetli Kromofob Renal Hücreli Karsinoma

ÖZET

52 yaşındaki bir kadında sarkomatöz komponentlere sahip kromofob renal hücreli kanserli nadir bir olgu sunmaktayız. Tümör, 70x60mm boyutunda, abdominal ultrasonla rastlantısal olarak bulundu. Abdominal bilgisayarlı tomografi sağ böbrek üst polden kaynaklanan sınırları belirgin solid bir tümör gösterdi. Histolojik olarak, tümör birbirine geçmiş iki farklı morfolojik komponentten ibaretti: kromofob renal hücreli kanser ve yüksek grade'li iğsi hücreli sarkom. Rabdoid hücrelerle karışmış oldukça atipik iğsi hücrelerden ibaret sarkomatoid bölge vardı. Bu tümörün eşsiz histolojik bulgusu kalsifikasyonun varlığı ve tümör boyunca psammoma yapı oluşumuydu. Bu olgunun kromofob renal hücreli karsinomdan sarkomatöz renal hücreli kansere bir tümör progresyon yolunun varlığını desteklediğini düşünmekteyiz. Kalsifikasyonlu sarkomatöz kromofob renal hücreli kanser üzerine literatür gözden geçirildi

Anahtar kelimeler: *Kromofob renal hücreli karsinom, sarkomatoid renal karsinom, kalsifikasyon*

INTRODUCTION

Sarcomatoid renal cell carcinoma represents 1-6.5% of renal cell carcinomas (1). The sarcomatoid differentiation is not a distinct histological entity and confers higher aggressiveness on any of the different subtypes of RCC (2,3). The incidence of sarcomatoid differentiation is 9% in chromophobe renal carcinoma (4). Renal cell carcinoma with sarcomatoid differentiation is a tumor with aggressive behavior. We report such a case and review the related literature.

CASE

52-year-old woman was admitted to our hospital with a complaint of vaginal bleeding. The patient had no symptoms related to a kidney tumor at diagnosis. The patient's past medical history included hypertension and diabetes mellitus. The urine analysis showed microscopic haematuria. Abdominal ultrasonography showed a leiomyoma of the uterus and incidentally the presence of a well circumscribed, heterogeneous solid lesion (70x65 mm) in the upper pole of right kidney. The tumor confirmed with abdominal computed tomography,

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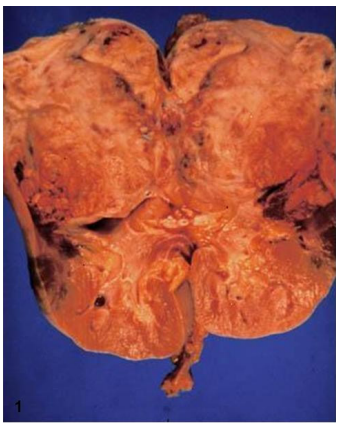

***Figure 1**. Macroscopic appearance of tumor which has circumscribed border toward the renal parenchyma and heterogenous.*

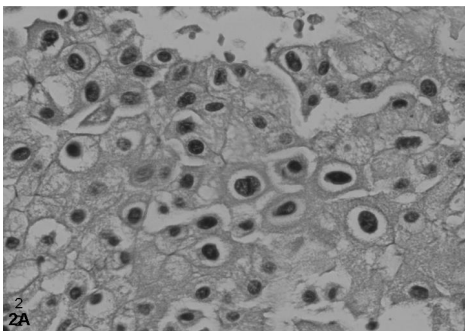

***Figure 2A**. The chromophobe renal cell carcinoma areas. The tumor cells with koilocytic appearance and accentuated cell borders (hematoxylin-eosin, magnification X200).*

and the patient underwent a right radical nephrectomy, retroperitoneal lymph node dissections, and total abdominal hysterectomy. The cut surface of the tumor had a variegated appearance, multilobulated, with white-grey, brown fleshy-appearing areas adjacent to the necrosis and hemorrhage with foci of dystrophic calcifications (Figure1). The tumor was limited to the kidney. Additional grossly unremarkable lymphs nodes were also present. Histologic analysis of the kidney revealed two distinct morphologic components of the tumor: a chromophobe renal cell carcinoma and high-grade spindle cell sarcoma. The chromophobe renal cell carcinoma consisted of compact epithelial cells arranged in a nested pattern and trabecular sheets. The cells were of medium size and had clear to granular cytoplasm with prominent cell borders. The centrally located nuclei were moderately pleomorphic, round to ovoid, with wrinkled nuclear membranes and finely clumped chromatin. Nuclei frequently displayed varying degrees of ''raisinoid'' changes. Nuclear halo was frequently seen in tumor cells. Occasional binucleated and multinucleated cells were identified. Single, prominent, small nucleoli were readily identified (Figure 2A). These histological features are characteristic of CRCC. The sarcomatoid component in this case was composed of areas resembling fibrosarcoma (Figure 2B) and malignant fibrous histiocytoma (Figure 2C). Nuclei were large, hyperchromatic, and showed frequent mitoses. Pleomorfism was marked. The tumor also contained focal rhabdomyosarcomatous component by round to polygonal cells with globular eosinophilic inclusion bodies in the cytoplasm, eccentric vesicular nucleus and prominent nucleolus (Figure 2D). The patient had only 10% to 15% of rhabdoid areas. Tumor necrosis and hemorrhage were frequently seen in the sarcomatous areas. The sarcomatoid portions of the tumor was composed less than carcinomatous component (less than 50% of the tumour volume). Venous invasion was observed in the case. No metastased was present in regional lymph nodes or to distant visceral sites at the time of diagnosis. Dissection of the uterus showed well-circumscribed, tan-pink, firm leiomyomas in the myometrium 10cm in dimension. No atypia or mitosis was noted. The remaining uterus, cervix, ovaries, and fallopian tubes were otherwise unremarkable. The patient died from widespread metastatic disease 12 months after nephrectomy.

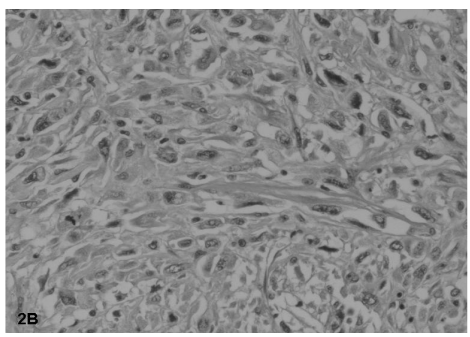

***Figure 2B**. Histologic findings of undifferentiated sarcoma-like features showing spindle cell proliferation, simulating a fibrosarcomatous pattern(hematoxylin-eosin, magnification X200).*

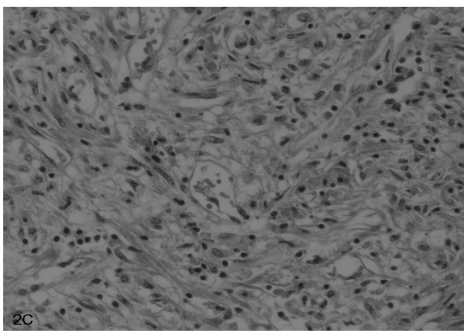

***Figure 2C**. Sarcomatoid area include pleomorphic plump spindle cells resembling malignant fibrous histiocytoma (hematoxylin-eosin, magnification X200).*

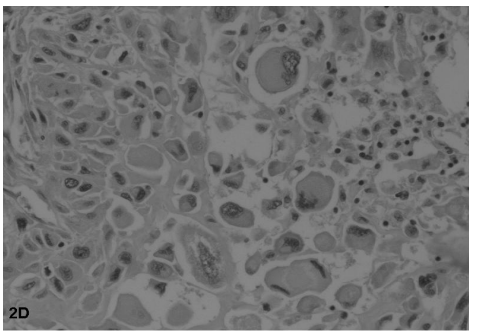

***Figure 2D**. Areas of large polygonal cells with eccentric oval atypical nuclei and eosinophilic intracytoplasmic inclusion resembling rhabdomyosarcoma (hematoxylin-eosin, magnification X400).*

DISCUSSION

Sarcomatoid renal cell carcinomas of the kidney are clinically aggressive tumors and are more commonly associated with clear cell renal cell carcinomas, but some authors suggest that the sarcomatoid tumor is most often associated with chromophobe RCC; however, the histological type does not influence the disease's outcome (5, 6). Both chromophobe carcinoma and sarcomatoid carcinoma of the kidney are rare (7, 8, 9). Identification of rhabdoid features as a morphologic variant of RCC is a recent report in few series (10, 11). Rhabdoid features are sometimes associated in RCC with sarcomatoid changes (12, 13). Renal cell carcinoma with sarcomatoid differentiation is a tumor with aggressive behavior that is poorly responsive to immunotherapy. In the series presented by Akkhtar et al (9), at the time of clinical presentation, all tumors exhibited an either locally aggressive growth pattern, with the sarcomatoid component extending beyond the renal capsule, or with involvement of the major vascular hilar structures. In our case, no metastased was present to regional lymph nodes or to distant visceral sites at the time of diagnosis. The proportion of sarcomatoid components may be an important factor. The prognosis of chromophobe cell carcinoma is believed to favourable however, an aggressive clinical course has been reported for tumors larger than 8 cm in diameter (14). In the present case, the tumor was 7 cm diameter. de Peralta-Venturina et al reported that the disease-specific survival rate of renal cell carcinoma with sarcomatoid change (22%), which is significantly lower than renal cell carcinoma without sarcomatoid change (79%) (5). In conclusion, we describe a case of chromophobe renal cell carcinoma showing sarcomatoid differentiation with the concomitant presence of extensive calcification which additionally had distinctive hyalinized, miksoid stroma and fibrosis. Increased awareness of this rare entity may improve its clinical and prognostic significance. Surgical resection may play role in the treatment of these cases.

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