Renal cell carcinoma sometimes exhibits 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 (1). 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 (1,2). Sarcomatoid transformation has been noted in virtually all types of RCC. However, chromophobe carcinoma is the most frequent renal cell carcinoma with a sarcomatoid component (2,3,4). We report an additional case and review cases in the literature of chromophobe renal cell carcinomas with a sarcomatoid component.

A 47-year-old woman presented with 12-kg weight loss and a history of right-sided abdominal pain, nausea, and vomiting. A physical examination revealed a cachectic woman with a large, firm, fixed and slightly tender right abdominal mass. CT revealed a right renal mass. A right radical nephrectomy was performed with no complications. There was disseminated tumor involvement in the right renal fossa and lymph nodes. The patient died 3 months after surgery.

Macroscopically, the resected mass was a well-circumscribed single mass, measuring 23x20x18 cm. The cut surface was gray tan in colour with soft areas extending through the renal capsule and distending the renal vein, and also had a firm consistency in some areas. The upper portion of the mass included kidney tissue measuring 6x4x2.5 cm.

Microscopically, the sarcomatoid component in this case was composed of intertwining bundles of spindle-shaped cells with various polygonal cells. The nuclei were large and hyperchromatic, and displayed frequent mitoses. The histological features of the sarcomatoid component varied from slide to slide. The sarcomatoid component was a high-grade fibrosarcoma in most areas, consisting of atypical spindle cells forming a storiform pattern (Figure 1A), but had more atypical, pleomorphic and plump spindle cells, simulating malignant fibrous histiocytoma in some areas. The chromophobe cell carcinoma consisted of large, round-to-polygonal cells with well-defined cell borders and amphophilic-to-pale-basophilic cytoplasm. Perinuclear halos were evident. The level of mitotic activity was very low (Figure 1B). In some areas, the sarcomatoid spindle-cell component intermixed with the chromophobe renal cell carcinoma (Figure 2A, B). In addition, the tumor exhibited geographic necrosis, calcification and ossification.

Immunohistochemically, the carcinomatous component exhibited strong diffuse positivity for epithelial
membrane antigen (EMA). Vimentin gave positive results only in the sarcomatoid areas. Neither ultrastructural studies nor staining with Hale's colloidal iron stain were performed.

Sarcomatoid renal cell carcinoma is an uncommon neoplasm consisting of both typical renal cell carcinoma and sarcomatoid components (5). This tumor has highly malignant biological behavior (1,5). Chromophobe carcinomas are the most frequent renal cell carcinomas associated with sarcomatoid components. Sarcomatoid chromophobe renal cell carcinomas do not exhibit any sex preference (2).

The diagnosis of chromophobe renal cell carcinoma in our patient was based on the usual features that characterize this tumor. The morphological appearance was that of a carcinoma composed of large epithelial cells, with well-defined cell borders and abundant reticular and translucent cytoplasm, and that of a fibrosarcoma composed of intertwining bundles of spindle-shaped cells.

The histological features of sarcomatoid renal cell carcinomas vary from case to case, and also in different areas of the same tumor (1). The sarcomatoid component of renal cell carcinoma may demonstrate histological features of fibrosarcoma or malignant fibrous
histiocytoma (1,6), and also may have osteosarcoma-like areas (7-10). The sarcomatoid components of our case generally consisted of high-grade fibrosarcoma areas. It has been reported that areas of hyalinized stroma interspersed with tumor nests are present in 41% of cases and are associated with smooth-muscle metaplasia in 26% of cases (11). In our case, we observed ossification within the sarcomatoid component with no smooth-muscle metaplasia.

Immunohistochemical and ultrastructural studies on the sarcomatoid component have demonstrated that in most cases the sarcomatoid cells are derived from the carcinoma cells. Chromophobe carcinomas have a greater propensity to transform into SRCCs than other types of renal cell carcinoma (2). The events responsible for the transformation of a chromophobe carcinoma cell into SRCC are not known exactly. The tendency for sarcomatoid transformation of chromophobe carcinomas is probably related to genetic make-up (3). However, there are some studies explaining this propensity. SRCCs arising in chromophobe cell carcinomas have a hypodiploid DNA pattern (2,3,12,13). Akhtar et al. (3) reported monosomy in chromosomes 1, 2, 6, 13, 17, and 21 in both chromophobe carcinoma and the sarcomatoid component. In addition, Oda et al. (14) demonstrated an extremely high mutational rate for the p53 gene in the sarcomatoid component but not in the carcinoma component. These findings indicate that the genetic pattern of the chromophobe carcinoma has a major role in the sarcomatoid component, and that there is a close association between p53 mutation and sarcomatoid transformation in RCC.

It is clear that additional studies are needed in order to understand the key genetic events that trigger transformation of chromophobe carcinoma to sarcomatoid carcinoma.

50% of SRCCs are derived from a background of chromophobe carcinoma (2). This indicates that there is a close relationship between chromophobe carcinomas and sarcomatoid transformation.

Ro et al. (1) reported that sarcomatoid renal cell carcinomas with zero or minimal necrosis yield a favorable prognosis, whereas those with moderate or massive necrosis yield a significantly poorer survival time. Our case displayed geographic necrosis in large areas. The proportion of sarcomatoid components also appeared to be an important prognostic factor. However, there is no correlation between tumor size and prognosis (1). Sarcomatoid renal cell carcinoma is a high-grade malignant renal tumor (2,4).

In conclusion, the tumor in our patient had high-grade fibrosarcoma and malignant fibrous histiocytoma-like areas together with ossification, and also showed a poor prognosis.

References