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Case Report

A rare and rapidly progressing renal chondrosarcoma: a case report

Tao He[1], Tianlei Kou[1], Xuyi Chen[2], Xuan He[1], Yun Guo[3]

Departments of [1]Pathology, [2]Research, [3]Endocrinology and Hematology, Characteristic Medical Center of The Chinese People's Armed Police Force, Tianjin, China

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Abstract: Mesenchymal chondrosarcoma of the kidney is a rare soft tissue sarcoma. We report a 64-year-old female with a right renal pelvic mass. After a right nephrectomy was performed, the histopathological examination confirmed the diagnosis to be primary extraskeletal mesenchymal chondrosarcoma. After the surgical removal of the tumor, the patient suffered rapid disease progression.

Keywords: Mesenchymal chondrosarcoma, extraskeletal mesenchymal chondrosarcoma, kidney, renal pelvis, sarcoma, primary renal malignant tumor

Introduction

Extraskeletal mesenchymal chondrosarcoma (EMCS) is a rare malignant neoplasm. Mesenchymal chondrosarcoma (MCS) is characterized by a biomorphic pattern that is composed of poorly differentiated small round cells and islands of well-differentiated hyaline cartilage [1]. MCS, which originally was thought to be restricted to bone, is reported in about one-third of these tumors occurring at extraskeletal sites [2, 3]. Most cases of EMCS occur in the meninges, the lower extremities, and the orbits [2], but a few can be occasionally seen in the parenchymal organs. Herein, we report a case of EMCS of the kidney in an elderly woman experiencing rapid disease progression.

Clinical features

The 64-year-old female patient was admitted to our hospital for urology due to a painless hematuria lasting for more than 20 days on February 25, 2019. The patient had no frequency or urgency of micturition. Flank pain and fever were absent.

The patient had several underlying chronic diseases. The patient underwent a cholecystectomy more than 10 years earlier and recovered well. The patient had a history of diabetes mellitus for more than one year, with a maximum fasting blood glucose of 8.6 mmol/L. The patient also had a history of coronary heart disease for more than 4 years and underwent a coronary angioplasty implant (CASI) 2 months earlier and was taking anticoagulant drugs such as aspirin. The patient had a history of smoking for more than 30 years, smoking an average of 10 cigarettes per day.

The laboratory findings were as follows: Red blood cells: $3.7*10^{12}$/L (3.8-5.1*10^{12}/L); hematocrit: 34.7% (35-45%); albumin: 38.9 g/L (40-55 g/L). Fasting blood glucose: 7 mmol/L (3.9-5.9 mmol/L); glycated hemoglobin: 6.5% (4.0-6.3%).

The abdominopelvic ultrasonography showed an unclear, hypoechoic solid mass (2.7 cm in diameter) located in the pelvis portion of the upper pole of the right kidney (**Figure 1A**). A cystoscopic examination of the right renal pelvis revealed a cauliflower-like mass, which was thought to be renal pelvic cancer (**Figure 1B**, **1C**). An abdominopelvic CT scan showed a mass with calcification in the upper part of the right kidney (the outer court).

Under the diagnosis of primary renal tumor, a transabdominal right radical nephrectomy was performed, and a sheet-like, well-circumscribed

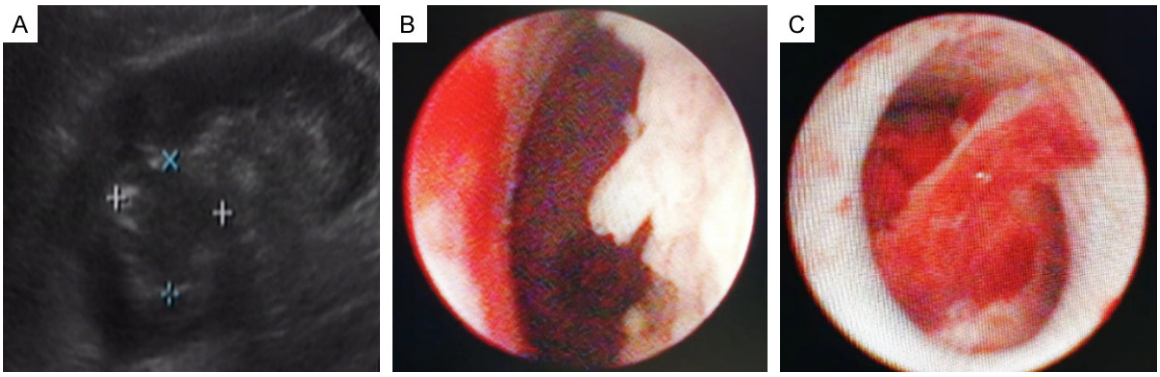

Figure 1. Abdominal ultrasound and cystoscopic examination of the right kidney. A. The abdominal ultrasound showed an unclear, hypoechoic solid mass (2.7 cm in diameter) located in the pelvic portion of the upper pole of the right kidney. B and C. The cystoscopic examination showed a cauliflower-like mass in the renal pelvis.

mass, 3.0 cm×2.5 cm, was found at the renal pelvis. The cut surface showed scattered cartilage foci in the gray-pink fish-like tumor tissue. Focal hemorrhage or necrosis was visible. Histopathology: The tumor consisted of poorly differentiated cells and scattered islets of cartilage. The poorly differentiated cells had a sheet-like distribution with scant cytoplasms. The nuclei were deeply biphasic stained, round, oval, or short spindle-shaped. The cartilage islands were relatively mature, variable in size, with calcification. Osteoclast-like, multinucleated giant cells occasionally could be seen (**Figure 2A-G**).

Immunohistochemical (IHC) staining showed: vimentin: positive, CD99: positive, S100: positive (**Figure 2H**), CD10: negative, CD34: negative, cytokeratin: negative, desmin: negative, WT1: negative, Ki67 (MIb-1): 20-30%.

The patient was discharged on March 30, 2019 and did not receive anti-tumor treatment such as radiotherapy or chemotherapy after her surgery. She was re-admitted to the hospital on May 16, 2019 and was admitted to the Department of Gastroenterology due to an incomplete intestinal obstruction. The thoracic CT showed multiple small pulmonary nodules, and metastases were considered. An abdominopelvic CT scan demonstrated multiple small nodules or swollen lymph nodes in the right peritoneum and pelvis, which were also considered metastatic. Clinical laboratory tests found that the serum ferritin (236.71, 4,63-204 ng/ml) was slightly increased. The patient's constitution was too poor to undergo chemotherapy, and her condition gradually worsened, and she ultimately died 3 months after the surgery.

Discussion

Mesenchymal chondrosarcoma (MCS) is a rare malignant soft tissue tumor. Until now, only 16 patients with renal MCS have been reported in the English literature since 1981 [4]. Renal MCS occurs in patients ranging in age from children to elderly people with the peak incidence in the third decade, with no significant gender differences. The manifestations of renal MCS are nonspecific, and flank pain and hematuria are two most common clinical symptoms. On clinical imaging, renal MCS is frequently misdiagnosed as renal cell carcinoma, renal urothelial carcinoma, or other rare malignant tumors. In our case, the kidney tumor was initially suspected to be renal urothelial carcinoma.

MCS is diagnosed mainly on the histopathological examination, with no specific Immunohistochemistry or molecular markers. MCS has typical biphasic pattern histological characteristics, which are composed of islets of well-differentiated cartilage surrounded by undifferentiated spindle mesenchymal cells, and it is not difficult to diagnose in typical cases. However, the amount of cartilage is highly variable. When the composition of the cartilage is rare, especially in fine needle aspiration specimens, the key for accurate diagnosis is to search carefully for the zones containing chondroid matrix. The differential diagnosis includes Ewing's tumor, hemangiopericytoma, synovial sarcoma, small cell osteosarcoma, and other small, round blue cell tumors [2]. Mesenchymal chondrosarcoma arising in the kidney can also be misdiagnosed as nephroblastoma (Wilms's tumor) [5]. Nephroblastoma characteristically contains undifferentiated blastemal cells, and some cases

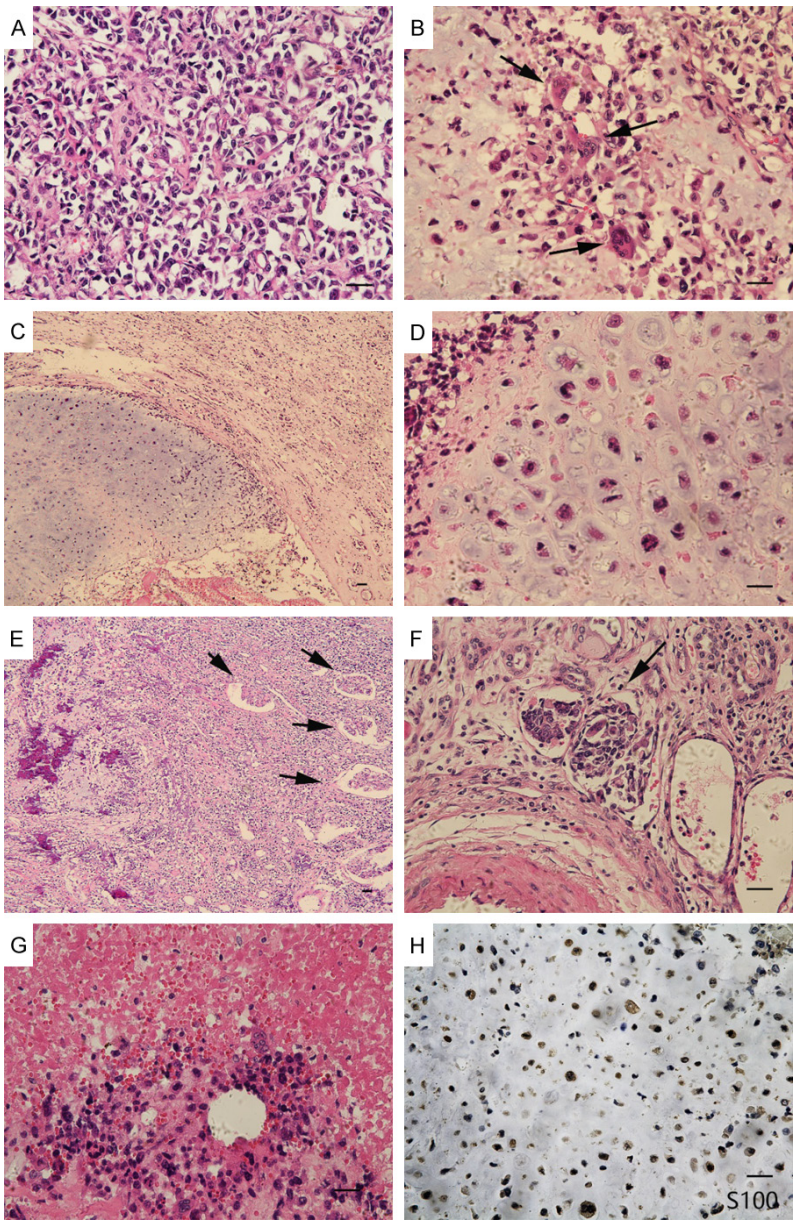

Figure 2. Microscopic and immunohistochemical examination of the renal extra skeletal mesenchymal chondrosarcoma. A. The area of the tumor containing poorly-differentiated, small, round to oval cells with scant cytoplasms (H&E, ×400). B. Area of the tumor containing an immature chondroid matrix and osteoclast-like multinucleated giant cells (H&E, ×400). Arrow: osteoclast-like multinucleated giant cells. C. Area of the tumor containing well-differentiated cartilage adjacent to the renal parenchyma (H&E, ×100). D. Well-differentiated cartilage surrounded by hyperchromic cells (H&E, ×400). E. Area of the tumor containing an osteoid-like matrix with calcification and glomerulus (H&E, ×100). Arrow: Glomerulus. F. Tumor emboli seen in the small blood vessels of the renal parenchyma (H&E, ×400). Arrow: Tumor emboli. G. Tumor necrosis (H&E, ×400). H. Immunohistochemistry showing S-100 positivity in the cartilaginous component (×400). Bar = 50 μm.

contain heterologous stromal differentiation including cartilage and bone. However, most cases of nephroblastoma occur in individuals aged < 10 years, and most of them contain a recognizable tubular or glomerular epithelial component, and 80% of the cases are immunoreactive for the WT1 protein [1].

MCS is an aggressive and rare neoplasm, and 15% of the patients are found with metastases at diagnosis [3], and distant metastases are observed even after a delay of > 20 years [2]. The correlation between the clinicopathological features and the prognosis of MCS is not clear. Some studies have shown that the hemangiopericytic features, the proliferation rate, and the extraskeletal origin are negatively correlated with the prognosis [6-8]; however, the conclusions are not universally accepted. In a retrospective study, metastases are found to strongly influence survival, and most of the patients will ultimately die from the disease. The median overall survival (OS) is about 3 years, and adjuvant chemotherapy is recommended [3]. In our case, the patient had extensive metastases two months after her surgery. Old age, underlying chronic diseases, and not undergoing systematic chemoradiotherapy are negatively correlated with survival.

Conclusion

We report a rare case of extraskeletal mesenchymal chondrosarcoma of the kidney with a rapid disease progression after surgery. The treatment for this primary renal malignant tumor with chronic

underlying disease in the elderly needs further optimization.

Acknowledgements

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Disclosure of conflict of interest

None.

Address correspondence to: Dr. Yun Guo, Department of Endocrinology and Hematology, Characteristic Medical Center of The Chinese People's Armed Police Force, 220 Chenglin Road, Dongli District, Tianjin 300162, China. Tel: +86-022-60578810; E-mail: 1982guoyun@163.com

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