Oncology

Pleomorphic undifferentiated sarcoma: A case of a giant renal mass

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1. Introduction

Renal cell cancers are the most prevalent type of malign renal masses. Sarcomas are one of the rarest subtypes. Primary renal sarcomas make up 1–3% of all renal malign masses. Previously known as malign fibrous histiocytoma (MFH), pleomorphic undifferentiated sarcoma (PUS) is the most prevalent soft tissue tumor in adults, with a 20% occurrence rate. PUS has a histologically wide range, the most prevalent forms being storiform and mixed form including pleomorphic areas. These tumors usually originate from renal parenchyma, and rarely from renal capsule.1 We present the diagnostic, therapeutic and pathological characteristics of a case of a giant renal PUS.

2. Case

83 year-old female patient applied to our clinic with nonspecific complaints such as abdominal pain, abdominal distention, loss of appetite, and weight loss. Abdomen was observed to be distended during physical examination. IV-contrasted full abdominal CT revealed a giant mass with 30 × 30 × 18 cm diameter, originating from the left kidney, pushing the kidney and the rest of the abdominal organs to the opposite side of the midline (Fig. 1).

The mass was heterogeneous, and neither necrosis nor calcification was observed, which increased the possibility of malignancy. Full body bone scintigraphy, Torax CT, and PET-CT were conducted for preoperative metastasis scans. There were not any metastasis observed in these tests. The patient was operated with radical nephrectomy, merging a supra-umbilical vertical midline and a left anterior subcostal incision. Left radical nephrectomy material weighted 7 kg, and was of a 30 × 28 × 20 cm size. On the surface of the section specimen, the mass was observed to suppress kidney tissue throughout the entire perirenal surface. The mass, beginning from the renal capsule and growing outwards, had large areas in fiber appearance, included solid areas, as well as loose myxoid areas cystic degenerated areas. A large cystic necrosis area was noted in the central (Fig. 2).

Regular-shaped adrenal gland was observed on the upper pole. In the microscopic evaluation, fibrohistiocytic cells with hemangiopericytomatic architecture were observed in H&E section specimens (Fig. 3A). There were findings of tumor infiltration including polygonal shaped bizarre nuclei with anaplastic appearance (Fig. 3B). In the cellular areas with marked atypicalness and pleomorphism, increased mitotic activity and large numbers of atypical mitosis were detected (Fig. 3C). Areas of bleeding and necrosis were noted. Immunohistochemical panel revealed widespread positive CD68-staining (Fig. 3D), focal positive CD10-staining, and focal weak SMA-staining. Pancytokeratin, HMB 45, S 100, desmin, CD34 and CD31 stainings were not detected. Histochemical evaluation of Ki67, proliferation index was about 20% (Fig. 3E), and mitosis was 10–17/10hf p. Significant in the lower pole, tumor was infiltrating to renal parenchyma, renal capsule, and perirenal fat tissue. Although the tumor was examined with numerous specimens, sarcomatoid renal cell carcinoma was excluded in the differential diagnosis because any carcinoma area was detected and any sarcomas were stained with pancytokeratin. Histo-morphological and immunohistochemical findings led to a report of pleomorphic indiffentiated sarcoma (malign fibrous histiocytoma; storiform-pleomorphic type).

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In order to prevent dehydration, hypovolemia, electrolyte imbalance, and hypovolemic shock in the postoperative early phase, the patient was monitored under intensive care conditions. Furthermore, intravenous liquid support was infused with balanced solutions, 150 cc/h. Beginning with the postoperative second day, the patient was monitored in the urology clinic, and there were not any complications. The patient is currently in postoperative third year, and there were not any recurrence or metastasis findings in the follow-up controls up to this point. Follow-up frequency was planned as every three months for the first two years, twice a year up to 5 years, and annually from then on.

![CT scan of the left kidney mass.](image)

**Fig. 1.** CT scan of the left kidney mass.

![Macroscopic examination of the tumor mass.](image)

**Fig. 2.** On macroscopic examination; heterogeneous giant tumor mass was dirty yellow in color and it located outside the lower pole of left kidney. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)
The first histogenesis of PUS tumors, previously known as MFH, is controversial. They were first defined in 1964, by O'bren and Stout. Tumor cells carry the indicators of both mesenchymal and mononuclear phagocytic system cells. In 1987, Ghandur-Mnaymanch suggested to rename it as fibrous histiocytic sarcoma, due to the behaviour of the tumor similar to sarcoma and lymphoma. After the recent change by World Health Organization, the term pleomorphic undifferentiated sarcoma was included in the literature, and malign fibrous histiocytoma was removed.

Until now, about 60 renal pleomorphic undifferentiated sarcoma have been published as case reports. These cases were almost equal in terms of sex and laterality. Most frequently observed symptoms are palpable abdominal tumor, fever, fatigue, weight loss and gastrointestinal problems. The diagnosis is completely based on pathology, and there are not any pathognomonic characteristics for radiological differential diagnosis. For this reason, they are usually hard to differentiate from renal cell carcinoma. The differentiation can be made using immunohistochemical staining. Similarly, our case applied to our clinic with nonspecific complaints such as abdominal pain, abdominal distention, loss of appetite, and weight loss and can only be diagnosed pathologically.

Best treatment option for undifferentiated renal sarcomas is complete surgical removal of the tumor. Initial plan should be total surgical excision, even though it is a tumor with high risk of local recurrence. We performed radical nephrectomy without any tumor on the surgical margin, so the prognosis of our case was favorable.
Therefore recurrence rate of PUS as 44%, lung (82%) and lymph node (32%) metastases is shown at high rates. Factors indicating negative prognosis include old age, large tumor size, depth of invasion, presence of tumor necrosis, high mitotic activity, invasion, and distant metastasis. Our case continues to her life despite the presence of all negative prognostic factors except distant metastasis. Any metastases or recurrences were observed within this 3-year period.

Due to local recurrence and distant metastasis, chemotherapy is often recommended with ifosfamide and doxorubicin. Furthermore, radiotherapy is recommended to tumor bed with positive surgical margin. In a 5-case series using cyclophosphamide, vincristine, Adriamycin, and actinomycine D regimen, was reported to no recurrence up to 2 years. General prognosis of undifferentiated renal sarcomas is poor, with a 14% survival rate for 5 years. Our case does not any adjuvant treatment for PUS, because the surgical margin was negative and the performance status of patient was too poor to be given any chemotherapy regimen.

4. Conclusion

A total surgical resection is sufficient to increase primary disease related survival period. Adjuvant chemotherapy is controversial for early phase and localised disease, and lacks significant support. Considering patient characteristics such as general condition and age, adjuvant chemotherapy may not be planned for the patient.

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Conflicts of interest

None.

Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.eucr.2018.01.002.

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