SARCOMATOID RENAL CELL CARCINOMA

DIAGNOSED BY PERCUTANEOUS NEEDLE BIOPSY

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ABSTRACT

A 56-year-old man presented with discomfort in the abdomen and backache. He lost his body weight by 10 Kg in 6 months.A large retroperitoneal mass was found in addition to the right renal mass.It seemed necessary to exclude additional tumor in theretroperitoneum.

Abdominal CT with and without contrast enhancement showed Tumors in the right kidney and giant tumors in the retroperitoneum. Percutaneous core needle biopsy of the retroperitoneal tumor and renal tumor was carried out under CT control.Pathological study revealed sarcomatoid renal cell carcinoma in the retroperitoneal mass, but mostly fibrous tissue in the renal mass.

Final diagnosis was sarcomatoid renal cell carcinoma with retroperitoneal extension. This type of renal cell carcinoma is noted to have very poor prognosis in the literature. Our case was also poor in prognosis, and the patient expired three months after the diagnosis was established.

Keywords : Sarcomatoid renal cell carcinoma, Renal cell carcinoma, needle biopsy, prognosis.

ABSTRAK

Intramuscular lipoma adalah kondisi yang jarang dan terhitung sekitar 1,8% dari semua tumor primer jaringan adiposa dan kurang dari 1% dari semua lipoma. Tumor ini berasal dari jaringan otot pada berbagai lokasi. *Giant intramuscular lipoma* otot biceps brachii adalah tumor yang jarang.

Seorang laki-laki usia 48 tahun dengan massa di lengan kanan atas. Massa tersebut muncul sejak satu tahun yang lalu dan ukurannya semakin bertambah besar. Pada pemeriksaan fisik, terasa nyeri pada saat palpasi dan *mobile*. Pada foto polos, tak tampak jelas massa *soft tissue* pada lengan atas. *Computed tomography* (CT) scan menunjukan massa hipodens pada otot biceps brachii dextra dengan densitas -72 hingga -83 Hounsfield. *Magnetic resonance imaging* (MRI), massa berasal dari otot biceps brachii. Pada T1 dan T2 weighted images, tampak lesi dengan

intensitas yang tinggi, dan SPAIR menunjukan densitas yang sama dengan lemak normal. Lesi tersebut di eksisi, didapatkan lesi dengan ukuran lebih dari 12 cm. hasil patologi menunjukan *intramuscular lipoma*.

Lipoma berukuran lebih dari 5 cm di klasifikasikan sebagai giant *lipoma*. Giant lipoma di ekstremitas atas, dari otot biceps brachii adalah jarang. Pada pemeriksaan foto polos, dapat tidak terlihat, atau tampak sebagai massa radiolusen dengan opasitas lemak. Pada CT dan MRI, lipoma tampak sebagai massa *non-invasive* dengan intensitas signal lemak yang homogen. Diagnosis banding utama intramuscular lipoma adalah liposarcoma berdiferensiasi baik. Penatalaksanaan yang dipilih adalah eksisi. Pemeriksaan patologi adalah pemeriksaan yang penting untuk konfirmasi diagnosis.

Kami laporkan kasus jarang *giant intramuscular lipoma biceps brachii* yang berhasil di eksisi dengan baik. CT dan MRI dapat mengidentifikasi dan melokalisasi tumor ini, dan memfasilitasi perencanaan operasi.

Kata kunci : intramuscular, lipoma, giant, biceps brachii

INTRODUCTION

A large retroperitoneal mass needs differential diagnosis between malignant lymphoma and metastases. A patient was referred to the Department of Urology because of a large retroperitoneal mass and suspected renal mass from the Department of Gastroenterology.

A needle biopsy was carried out and diagnosis of sarcomatoid renal cell carcinoma was established by pathological study. Herein, we report the case and review the literatures of sarcomatoid renal cell carcinoma.

CASE REPORT

A 56-year-old man presented with discomfort in the abdomen and backache. He lost his body weight by 10 Kg in 6 months. Physical examination revealed slight tenderness in the epigastrium and right lower quadrant of the abdomen. The blood pressure was 121/66 mmHg. Pulse rate was 67/min.

The chest x-ray showed a mass in the left lower lung field, suggesting metastasis. Plain film of the abdomen showed a large mass in the lower pole of the right kidney. Abdominal plain CT showed tumors in the right kidney and giant tumors in the retroperitoneum.

Contrast-Enhanced CT showed a large renal tumor in the inferior and posterior portion of the right kidney and a large mass in the retroperitoneum adjacent to the right kidney and extending to the anterior portion of the abdominal aorta. There were also the right para-renal mass and para-aortic lymph node swelling. The renal tumor was irregularly enhanced, but the retroperitoneal mass was not stained significantly. The renal tumor was consistent with renal cell carcinoma (RCC), but the pathology of the retroperitoneal

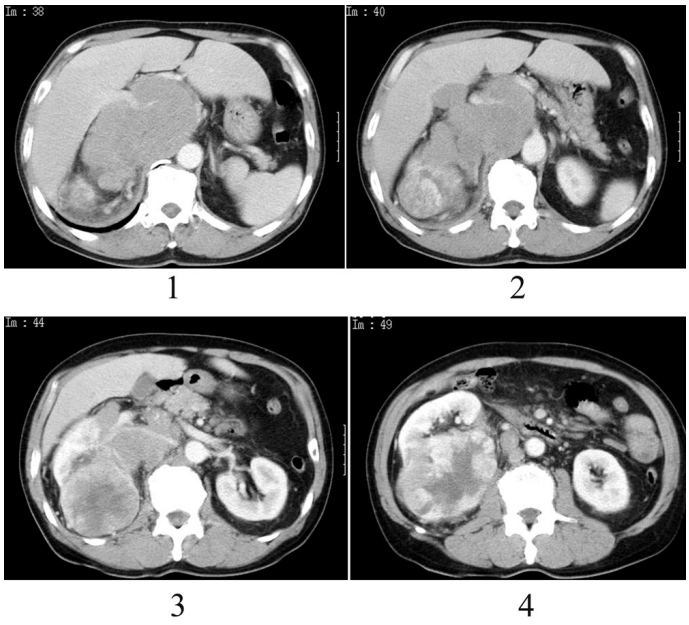

Fig. 1. Enhanced CT of the tumors in the right kidney and in the retroperitoneum.
Four slices from the upper part of the tumor to the lower portion of the right kidney (1 to 4). The renal tumor was irregularly enhanced, but the retroperitoneal tumor was only faintly enhanced.

mass was not certain. (Fig. 1)

The possible differential diagnoses were 1. Renal cell carcinoma with extensive metastases in the retroperitoneum, 2. Concurrent renal cell carcinoma and malignant lymphoma, or 3. Both tumors were malignant lymphoma.

Laboratory data are as follows:

WBC 17,930, RBC 440 x 10^4, Hb 13.3, Plt 31.5, TP 8.2, T Bil 0.44, BUN 12.8, Cr 0.70, AST 14, ALT 20, ALP 354, CRP 2.65, LDH 241, GLU 88, SCC Antigen < 1.0, CA 19-9 22, CEA 0.8, S-AMY 47, Electrolytes WNL

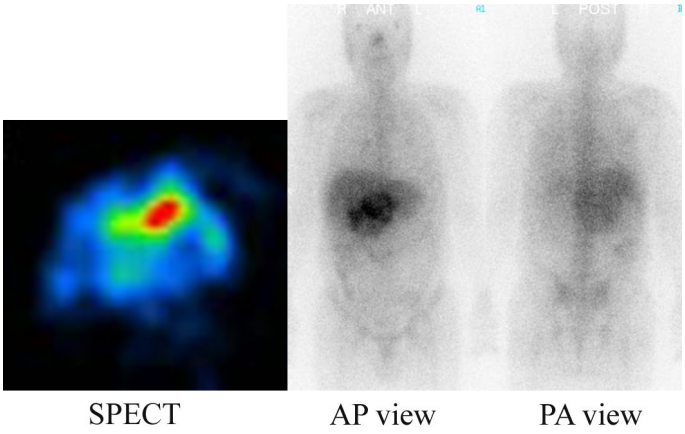

Fig. 2. Ga Scintigraphy
111MBq of Ga citrate was injected. The frontal view (AP view) shows high uptake in the upper part of the tumor. Posterior view (PA view) shows less marked uptake in the right upper quadrant of the abdomen. SPECT shows the highest uptake in the upper and left side of the mass in the slice section.

Study by nuclear medicine:

A Ga scintigraphy was carried out by the injection of 111 MBq of Ga citrate, which showed a high uptake in the retroperitoneal tumor, but much less uptake in the right kidney. (Fig. 2). Therefore, retroperitoneal malignant lymphoma was also entertained as a differential diagnosis.

A plain MRI study of the abdomen was added. The coronal images of MRI showed a large mass in the retroperitoneum as well as a large mass in the lower portion of the right kidney and also a smaller mass in the upper pole of the kidney. The smaller mass in the right kidney on the previous axial CT seems to have been obscured by the retroperitoneal giant mass. (Fig. 3)

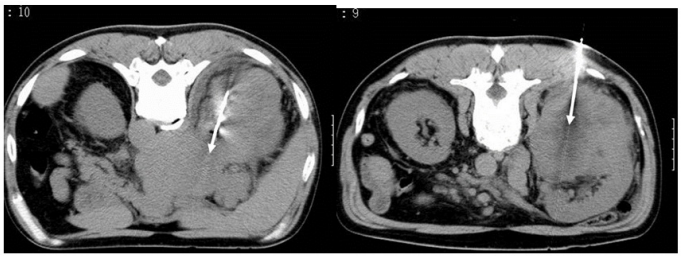

Fig. 4. Needle Biopsy of Retroperitoneal & Renal tumors.
Core needle biopsies were carried out under CT control. CT shows the positions of the tips of the biopsy sites.
The periphery of the renal tumor was not biopsied (B), but it seems to be necessary to make the correct diagnosis of renal tumor, although we obtained correct diagnosis of sarcomatoid renal cell carcinoma from the tissue in the retroperitoneal mass (A).

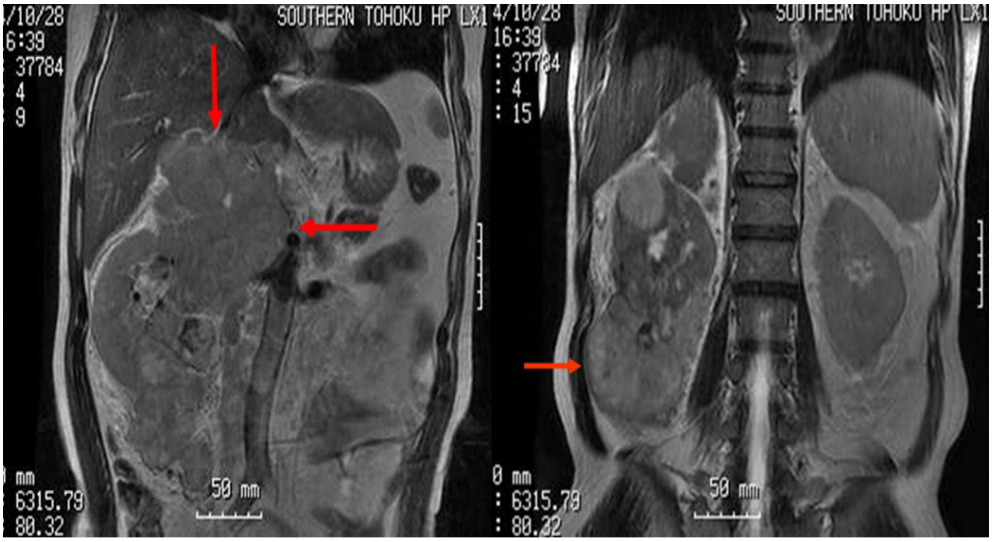

Fig. 3. MRI of the kidney and retroperitoneum. (T2 WI, Coronal images) MR images show a large mass in the upper abdomen adjacent to the right kidney (A), and lower pole of the right kidney and a smaller mass in the upper pole of the right kidney (B).

Pathological study by needle biopsy:

A core needle biopsy of the retroperitoneal and renal masses was carried out and the tissues obtained were studied pathologically.

A Super-Core II, Disposable Semiautomatic System (Sheenman Co. Ltd.) was used. The portions of biopsy were depicted by localization CT in the prone position. (Fig. 4)

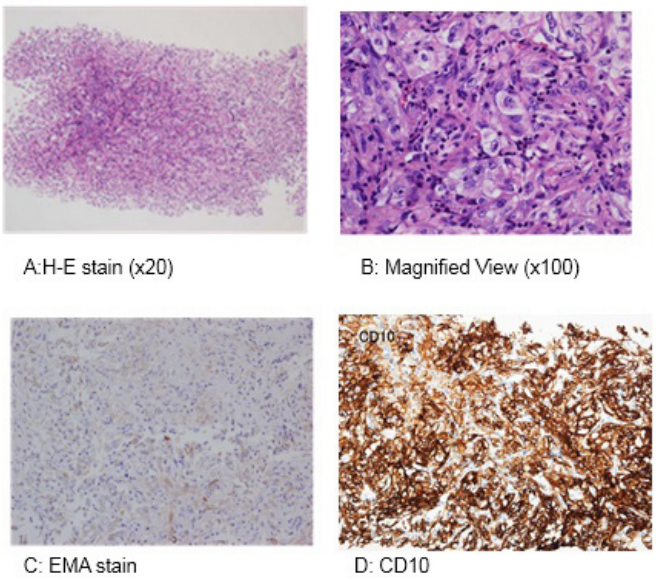

Fig. 5. Photomicrographs and immuno histochemical studies of the biopsy tissues from the retroperitoneal mass.
A: H-E stain (x20):Photomicrograph shows short round to spindle tumor cells, which are irregularly arranged.
B: Magnified view of H-E stain (x100): Photomicrograph shows the tumor cells to have clear to eosinophilic cytoplasm and large and small ovoid or irregular nuclei with distinct nucleoli. Multiple nuclei are also noted. Tumor cells are gathered densely. Mitoses are scattered.
C: EMA stain is positive, revealing cell membrane element.
D: CD10 is positive.

Final pathological diagnosis was renal cell carcinoma, grade 3, sarcomatoid type.

Pathological study revealed renal cell carcinoma, grade 3,sarcomatoid type in the tissue from the retroperitoneal tumor.No lymphatic tissue was present. Hematoxyline-eosine stain (H-E stain) showed short spindle tumor cells, which were arranged irregularly. Magnified H-E stain showed the tumor cells to have clear to eosinophilic cytoplasm and large ovoid or irregular nuclei with distinct nucleoli. Immunohistochemistry of the tissues showed positive EMA and CD10. (Fig. 5)

The tissue from the right kidney yielded mostly fibrous tissue and no tumor tissue was found. Pathological diagnosis was the right sarcomatoid renal cell carcinoma with retroperitoneal extension.

The patient was placed on IL-2 therapy (700,000 unit of IL-2 in 500 ml normal saline). He was once discharged from the hospital, but he was readmitted when he developed obstructive jaundice by enlarged retroperitoneal tumor and general malaise, and he expired three months after the diagnosis was established.

DISCUSSION

Sarcomatoid renal cell carcinoma is a rare type of renal cell carcinoma, and it was found only 37 cases among 2100 cases (1.8%) of renal cell carcinoma at the Mayo Clinic.

Bennington and Beckwith[1] describe that most common forms of such lesions were those mimicking Rhabdomyosarcoma (10 cases) and fibrosarcoma (24 cases). In many of the sarcomatoid forms, characteristic patterns of renal adenocarcinoma may be found if enough sections of the tumor are examined.

The majority of tumors classified as renal sarcoma reviewed by the authors have proved to be sarcomatoid renal adenocarcinoma by histochemical technics and careful examination of multiple sections. Electron microscopy: Sarcomatoid renal adenocarcinomas do retain certain features of epithelial cells which help in distinguishing them from sarcoma (Tannennbaum).

Cangiano et al. retrospectively reviewed 31 consecutive cases of sarcomatoid RCC during 1990 – 1997 at UCLA[2] They found metastases at the time of radical nephrectomy in 84% of cases. Metastases were in the lung (67%), bone (40%), liver (21%), lymphatics (33%) and brain (15%).

The patients were placed o Immunotherapy in 81% of cases, with interleukin (IL)-2-based therapy, and others. One- and 2-year overall survival rates were 48% and 37%, respectively. They concluded that surgical resection and high dose IL 2-based immunotherapy may play a role in the treatment of sacomatoid RCC in select patients.

Wu and his associates[3] reported a case of sarcomatoid renal cell carcinoma. Their case was a 42-year-old male patient presented with lower leg edema, shortness of breath, body-weight loss of 5 kg over a 2-month period. Physical examination revealed the patient to be pale, of cachexia, and palpable abdominal mass in right abdomen. Enhanced CT revealed a giant renal tumor on the right, measuring about

28 cm in diameter, extending upwards, backwards and also to the left side of the abdomen. The tumor was irregularly enhanced peripherally with relative low density inside. Biopsy of the right kidney confirmed sarcomatoid RCC. They reported that sarcomatoid RCC is an aggressive tumor and its prognosis was poor.

Shuch and his associates[4] states in their comprehensive review that although accounting for only 5% or so of renal cell carcinoma, the aggressive nature and advanced stage of presentation makes sarcomatoid renal cell carcinoma fairly common to practitioners who manage patients with metastatic disease. They also introduced the pathology of sarcomatoid RCC as many urologists and medical oncologists consider sarcomatoid RCC to be a clinically relevant grouping, and the histology contains features similar to sarcomas, with spindle-like cells, high cellularity, and cellular atypia; necrosis and microvascular invasion are present in 90% and 30 % of cases, respectively; the majority of tumors have a variable amount of recognizable carcinoma elements.

They further states that in clinical presentation sarcomatoid RCCs are usually extremely large, with a mean tumor size of 9-10 cm. The incidence of metastatic disease is extremely high at presentation, with 45-84% of cases; location of metastases are lung, bone, nodes, liver and brain. They states that core biopsy has emerged as a safe and reliable way of identifying renal malignancy and may replace fine needle biopsy. We also succeeded in making diagnosis of sarcomatoid RCC by core biopsy.

The median survival time of sarcomatoid RCC reported by majority of institutions is only 4-9 months after diagnosis. As to the treatment of sarcomatoid RCC, Shuch and his associates concluded as "There may be a role for combination chemotherapy with antiangiogenetic therapy in sarcomatoid RCC, but the ultimate improvement will come from better molecular and genetic characterization of sarcomatoid RCC and design of specific therapies."

According to Stratton and his associates[5] cancer genomes have been studied extensively and approximately 100, 000 somatic mutations from cancer genomes have been reported. Simplified technology to assess alteration of genes is rapidly advancing recently.

Santarpia and his associates[6] described that targeted drugs in small-cell lung cancer is limited in its application and express a hope in immunotherapy. However, targeted drugs for lung small cell carcinoma seem to be effective in our experience in selective patients with marked prolongation of the surviving time, when genomic characterization was confirmed to be suitable for the treatment of the specific cancer. Genomic drugs and immunotherapy will be further developed, and it is hoped that we will have a better method of treatment for sarcomatoid renal cell carcinoma also.

Regarding biopsy, the true pathological diagnosis depends on the correct site of tumor tissue without necrosis or marked degeneration. The tissue obtained from the renal tumor in our case was from the center of the tumor and not from the periphery of the tumor. It would be assumed that we could have obtained the diagnosis of sarcomatoid RCC, if we biopsied the peripheral portion of the renal tumor.

CONCLUSION

A percutaneous needle biopsy is a useful method to establish pathological diagnosis of tumors. We reported a case of sarcomatoid renal cell carcinoma, a rare type of RCC by core needle biopsy, and its poor prognosis of the disease. A few literatures on sarcomatoid RCC were reviewed.

Some parts of this article were reported at the 21th Annual meeting of Northern Chapter of Japanese Society of Interventional Radiology on July 19, 2008, and also at the 12th Annual Scientific Meeting of Indonesian Society of Radiology in Yogyakarta on May 6, 2017 as a plenary lecture: "Less invasive treatment for cancer patients by Interventional Radiology" by Shoichi D. Takekawa.

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