

## Case Report

# A case report of huge perirenal liposarcoma associated with renal cell carcinoma and reviews of three previous cases

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Received August 28, 2014; Accepted October 23, 2014; Epub November 15, 2014; Published November 30, 2014

**Abstract:** Retroperitoneal liposarcoma (RPLS) is a rare malignant tumor yet with a high recurrence rate. It is mostly observed in limbs and during retroperitoneal clearance, and accounts for nearly 40% of all adult retroperitoneal soft tissue sarcomas [1]. Renal cell carcinoma (RCC) is a common cancer developed in urinary system and its occurrence rate accounts for 3% of whole body malignant tumors [2]. In this report, we present a case of large perirenal liposarcoma associated with RCC, which is to date the fourth identified case. We analyzed the clinical pathologic data of this patient and reviewed the other three cases. Compared to the other cases, this patient is diagnosed at the youngest age and the liposarcoma tumor is with the largest diameter. Moreover, we have been following up the patient after operation for the longest time. All of these made this report rare and important.

**Keywords:** Retroperitoneal liposarcoma, tumor, case report

## Introduction

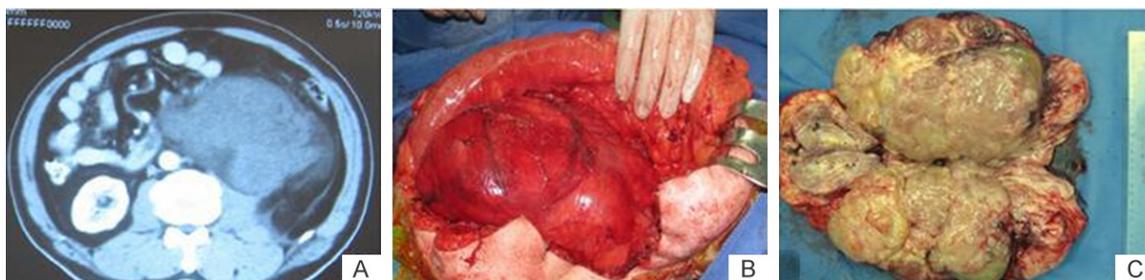
Huge perirenal liposarcoma is rarely observed clinically, and the renal cell carcinoma accompanied by RPLC is even more unusual. According to all published medical literature, there are so far only four cases including the patient reported here. And each of the four cases showed its own characteristics, which will be discussed below

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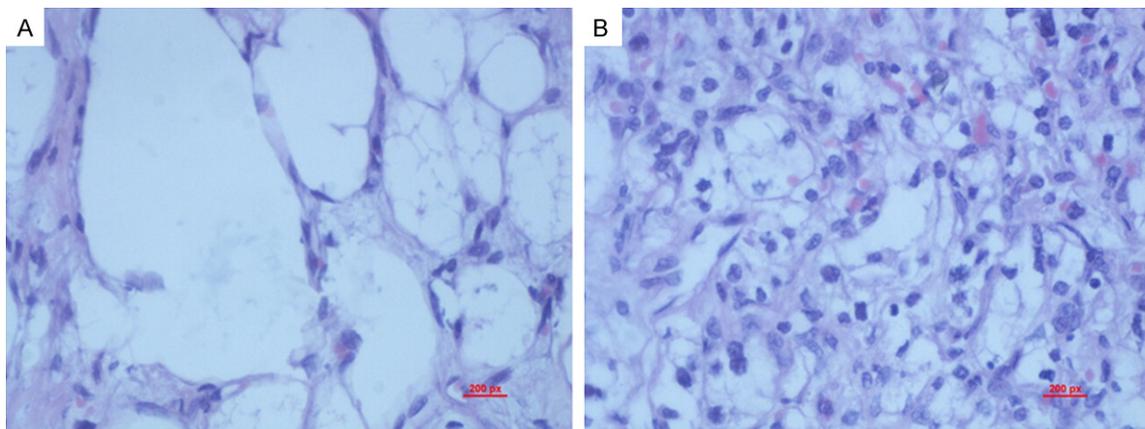
A 55-year-old male patient suffered from abdominal pain and abdominal distension for half a month. A large abdominal mass was noted on the left abdomen without clear causes. After seven days, the symptoms were aggravated and the abdominal mass become bigger prior to his admission to the hospital on July 14th 2010. On body examination, the patient was found to experience left abdominal dilation, and a large, solid abdominal mass extended from the left side of the rib arch to the symphysis pubis. The top was located on the costal margin, the bottom in pelvic cavity, dot-

ted inside, and separated into many parts. Left kidney is 12.8 cm × 7.2 cm, with 7.8 cm × 4.4 cm liquidity separation inside. Hint: cystic mass in the left abdomen, left hydronephrosis. Abdominal enhanced computed tomography (CT): large soft tissue mass, 35 cm × 20 cm × 15 cm, was observed on the left side of the renal hilum, CT 18, low density in the diseased area-48. Peripheral vascular and organ changed location under pressure. The strengthened reinforcement is not obvious, with mild delay to strengthen. It may be because of the left kidney intra-fascial placeholders and liposarcoma gets the highest possibility (**Figure 1A**). Kidney ECT showed that left kidney moved up and shrank, no abnormality was observed in blood perfusion, renal parenchymal damage, or delayed excretion. The patient underwent the resection of the tumor combined with left nephrectomy. During surgery, we observed that the mass located inferior of spleen in the pelvic cavity and from the left side of the abdominal cavity to completely encompass the aorta abdominalis. The mass invade into left kidney, left ureter and the left side of the psoas major (**Figure 1B**). The removed tissue weighed about

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**Figure 1.** A: Preoperative enhancement CT; B: Operation picture; C: Complete resection of the tumor.



**Figure 2.** A: Retroperitoneal liposarcoma observed under microscope: tumor are formed by spindle cells and adipose tissue with the shape of Beam pattern fiber. Slightly larger Spindle nuclei and it is with some atypia. Blood vessels are rich in mesenchyme with varying degrees of myxoid degeneration (H&E staining  $10 \times 40$  times). B: Renal clear cell carcinoma observed under microscope: Tumor cells show a funicular or gland bubble distribution, and cells were polygonal, circular center, nucleolus and abundant in cytoplasm (H&E staining  $10 \times 40$  times).

5.1 kg, with the size of  $40 \text{ cm} \times 25 \text{ cm}$  (**Figure 1C**). After the operation, we followed up for 34 months and the patient remain well without detectable recurrence of tumors. The tumor report was atypical lipomatous tumour (spindle cell subtype) with human renal clear cell carcinoma (**Figure 2**). IHC in removal of liposarcoma exhibited a vimentin<sup>+</sup> NF nucleus<sup>+</sup> C D68<sup>+</sup> CD34<sup>+</sup> phenotype. Resection tissue from kidney were CK<sup>+</sup> CD10<sup>+</sup>.

### Discussion

Here, we reported a patient with an enormous perirenal liposarcoma associated with renal cell carcinoma. According to published literature, it is the fourth report of such symptoms. Among the other three cases, the first one was perirenal liposarcoma containing extramedullary hematopoiesis associated with renal cell carcinoma observed during kidney cancer treatment [3]. The second was an incidental

finding of renal cell carcinoma in recurrent retroperitoneal liposarcoma [4]. The third one was discovered in the specimen of an operation [5]. Interestingly, all of the four cases were male patients with an average age of 66.2. The patient reported here was the youngest with the largest perirenal liposarcoma of  $40 \text{ cm} \times 25 \text{ cm}$ . And after the proper operation on the patient, so far, we have followed up for 34 months (**Table 1**).

RPLS is often observed in adult men, at the age from 40 to 70. It is a malignant tumor with slow growth rate. RPLS is hard to be identified at an early stage and 80% of the patients suffer an abdominal mass initially [6]. They may also suffer digestive tract symptoms such as abdominal distension and abdominal pain; or lower limb swelling and the difficulty in urination and defecation; or local compression symptoms and chronic consumption symptoms such as fever, fatigue, and thinness. Occasional acute

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**Table 1.** Reported cases of perirenal liposarcoma associated with renal cell carcinoma

First author	Year of publication	No. of cases	Age/ gender	liposarcoma size	treatment	Outcome (months)
Donald J	1994	1	71/M	No data	right nephrectomy	17
Yoshiaki K	2007	1	60/M	7.6 cm × 5.0 cm	right nephrectomy	24
Williamson JML	2008	1	79/M	40 cm × 20 cm	Re-operation	no data
Present case		1	55/M	40 cm × 25 cm	liposarcoma resection With nephrectomy	34

abdominal pain caused by tumor rupture, hemorrhage, and shock were also reported [7]. Typical RCC symptoms include blood urine, abdominal pain, renal area pain and abdominal mass. Other systematic symptoms include weight loss, fever, night sweats, decreased appetite, fatigue, etc, which are all common symptoms of malignant tumors [8]. Moreover, cancer metastasis directly causes symptoms such as chronic cough resulted from pulmonary metastasis, bone pain due to bone metastases; while transfer of organs such as liver metastases can cause jaundice symptoms. Occasionally visible lower limb swelling caused by tumor compression and varicocele can be observed [9]. Of all four cases reported, abdominal distension was the shared early symptom. Progressive increase of abdominal mass was observed in 1 case, diarrhea with angular cheilitis in 1 case, while no obvious symptoms in the other 2 cases. However, visible gastrointestinal symptoms in patients with urinary tract symptoms are often not obvious, thus it is prone to be misdiagnosed or not diagnosed.

Clinical diagnosis of RPLS or RCC mainly relies on imaging examination, for example, ultrasound (US) and CT examination are the most commonly used methods. US can display the location, size, type (solid or cystic), and number of tumors. It is non-invasive, cheap, and widely used in preoperative examination and postoperative review. CT has high accuracy in positioning and of great value in differential diagnosis of RPLS, thus is recommended for preoperative examination. RCC examined by magnetic resonance imaging (MRI) shows the symptoms of renal contour abnormalities, local boundary missing, cortex and medulla, compression of adjacent renal pelvis and calyces on formation and tumor pseudocapsule characteristic, etc. Intravenous urinary tract imaging and urinary tract imaging can detect the size of the renal tumor, location, and degree of invasion of col-

lection system. Ultrasound and CT examination were performed in all four cases, MRI in 2 cases. Thus, visible ultrasound, CT or MRI and other imaging methods should be the main diagnostic tools in the identification of RPLS with RCC. Histologic subtypes of liposarcoma include polymorphism, round cell, mucous, differentiation and dedifferentiation. 2 cases were differentiated liposarcoma, while the other two cases were pleomorphic liposarcoma. 2 cases were renal clear cell carcinoma, 1 case was renal papillary carcinoma, and 1 case was renal medullary carcinoma. Thus, pathological classification of visible RPLS with RCC is not definite.

Treatment of RPLS emphasizes the thoroughness and integrity. Tumor-adjacent fat tissue should be removed thoroughly, while the retroperitoneal lymph nodes should be cleaned as much as possible. As there is no complete capsule, tumor growth is rapid. Easily invading neighboring large blood vessels and the surrounding tissue organs, once the invasion, if conditions permit patients' tolerance, one-time combined resection of multiple organs or parts of viscera, tumors were resected totally in order to achieve the purpose of radical. Radical nephrectomy therapy on RCC is still the golden standard. Intraoperative resection of most kidney, ureter and renal week fascia, fat and kidney door surrounding lymph nodes, if affected adrenal, also should be firstly removed. Of all 4 cases, 2 patients underwent combined resection of retroperitoneal tumors and the left kidney, 2 underwent radical resection of the right kidney, indicating that radical resection is the main method of treatment of RPLS with RCC. Due to lack of randomized controlled study, curative effects of postoperative radiotherapy and neoadjuvant chemotherapy are unable to be evaluated and remain unknown. So far, 21 times of postoperative chemotherapy, with postoperative radiation therapy have been

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applied to the patient reported here, who is generally in good condition, showing no tumor recurrence and metastasis. Though notably, postoperative radiotherapy is revealed to be curative for RPLS with RCC, further research needs to be done to draw conclusions with statistical significance. To sum up, abdominal signs, ultrasound, CT and MRI imaging examination are the main diagnostic criteria of RPLS and RCC. Pathological classification is not standardized. Radical resection is the major method of treatment for this type of disease and the effect of postoperative radiotherapy is good. Because of the few reported cases of the disease, pathogenesis and treatment methods are yet remain further discussed. In the case reported in 1994, fat tissue was found of large amount and renal surrounding and hematopoietic tissue, and the well-differentiated liposarcoma with medullary hematopoiesis was also observed. This case was failed to diagnose kidney cancer, but preoperative renal function and intraoperative were found. During the surgery we found it is hard to keep the kidney and we did the joint resection. Therefore, it is rather important to conduct preoperative diagnosis and evaluations for this kind of patients. Otherwise it can easily cause the uncompleted surgical operation and may lead to mortality.

### Acknowledgements

Thanks all authors.

### Disclosure of conflict of interest

None.

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