Case Report

A Case Report of Ruptured Malignant Neuroendocrine Tumor of Kidney, an Extraordinary Presentation

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We report the case of a 51-year-old Thai female presented with right abdominal mass with pain and anemia. The radiographic findings showed ruptured renal mass. She underwent radical nephrectomy and the pathological report was malignant neuroendocrine tumor. Palliative chemotherapy was administered. The patient passed away three months after the operation. Renal neuroendocrine tumors are rare. Moreover, ruptured renal tumors are very rare. They are usually considered to be angiomyolipoma or renal cell carcinoma. Poor differentiation and rupture at presentation are aggressive features of tumors. To the best of our knowledge, this is the first case report of ruptured renal malignant neuroendocrine tumor.

Keywords: *Kidney neoplasms, Neuroendocrine tumors, Rupture*

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According to the 2004 WHO classification, neuroendocrine tumor is one of tumors of kidneys[1]. The incidence is less than 1% of tumors found in this organ[2]. The cellular origin remains unclear[3-5]. Moreover, ruptured renal tumors are not commonly seen. They are usually considered to be angiomyolipoma or renal cell carcinoma[6]. We report a case presented with ruptured neuroendocrine carcinoma of kidney.

Case Report

A 51-year-old Thai female presented to the emergency department with acute right upper quadrant pain for 2 hours. She detected abdominal mass at her right upper abdomen for 3 months. It progressively enlarged without pain. She had no jaundice, abnormal stool or hematuria. She lost her weight eight kilograms in one month. She had no symptoms of episodic headache, palpations or sweating. Three days ago, she gradually got pain at her mass without history of trauma. She noticed that the mass slightly increased in size. On physical examination, her vital signs were stable. She was mildly pale. Her abdomen was mildly distended with tenderness at right upper quadrant mass. The mass was about 20 cm in diameter. No rebound tenderness was detected. Her hematocrit was 21.6%. She was sent for computed-tomography (CT) scan. The CT scan showed a large complex solid-cystic mass from right kidney, about 14.6x18.2x25.0 cm (Fig. 1). High density clot anterior aspect of the renal mass below to lower margin of liver could be sentinel clot. Hemoperitoneum was suspected from generalized high density fluid in abdominal and pelvic cavity. A 1.0 cm arterial enhancing lesion at hepatic segment 2 was

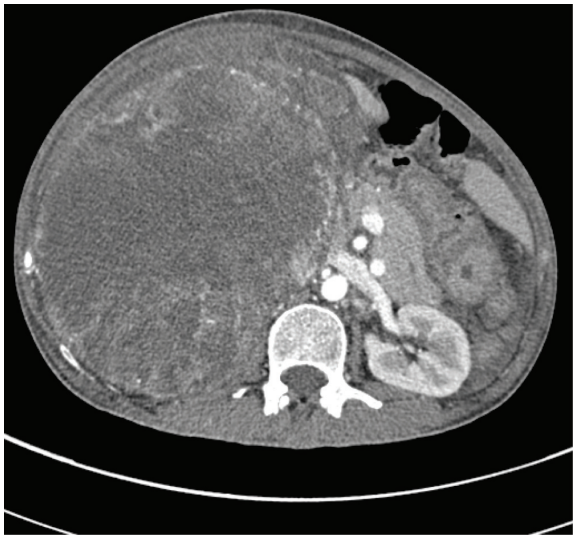

Fig. 1 Abdominal computed-tomographic image with contrast media showed huge complexed solid-cystic mass with evidence of intralesional hemorrhage which originated from right kidney.

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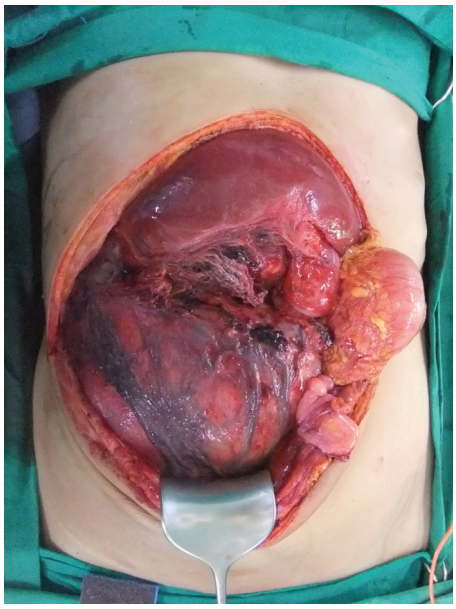

Fig. 2 Ruptured tumor at upper pole of right kidney.

suspected to be liver metastasis. At that time, the diagnosis was ruptured right renal mass with hemoperitoneum. The patient was resuscitated and three units of packed red cell were infused. Her hematocrit was stable about 30% and her abdominal sign was not progressed. Right radical nephrectomy with liver wedge resection was performed. Intra-operative finding was ruptured tumor at upper pole of right kidney (Fig. 2). The tumor adhered to right lobe of liver. Hemoperitoneum was found about 700 ml. Multiple small (less than 1 cm) intra-abdominal lymphadenopathy was detected.

The pathological report was an enlarged kidney with frequent necrosis and abundant fibrinous exudate covering the external surface, weighing 1,034 g. Bisection of the kidney revealed a ruptured poorly circumscribed tumor, involving almost the entire kidney, measuring 25x15x9 cm. Histologically, the preserved part of the tumor showed marked hypercellular (Fig. 3). The neoplastic cells possessed large, ovoid to fusiform, hyperchromatic nuclei with non-contiuous (stippled) chromatin, and inconspicuous nucleoli. The cytoplasm was indistinct. They were immunohistochemically reactive with chromogranin A (diffuse and intense) (Fig. 4) and synaptophysin (focal but intense). They did not show positive immunoreactivity for AE1/AE3, vimentin, RCC, CD10, SMA, S-100 protein, CD117, myogenin, desmin, and HMB45. No metastatic carcinoma in the regional lymph nodes was seen. Liver margin and liver segment 2 found no tumor cell.

After the recovery period, oncologist was consulted and palliative chemotherapy (carboplatin and etoposide) was administered. Two months later, the patient had abdominal discomfort and her abdomen was distended. CT scan was done. The study revealed multiple hepatic metastases, multiple intraperitoneal lymphadenopathy, and carcinomatosis peritonei. Chest X-ray showed new pulmonary nodules. Three months after the operation, she developed dyspnea, respiratory failure and passed away.

Discussion

Renal neuroendocrine tumor is an extremely rare disease, less than 1% of all epithelial renal

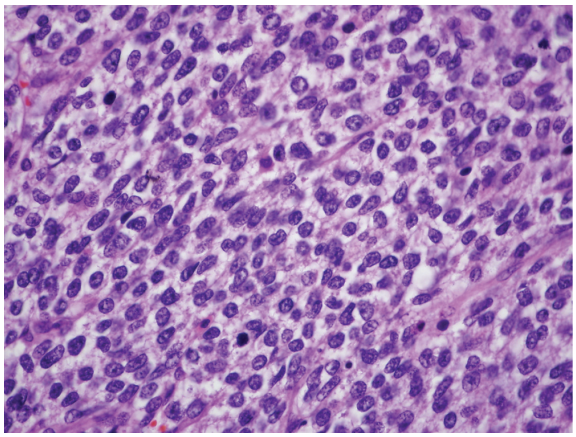

Fig. 3 Hypercellular tumor with hyperchromatic nuclei, stippled chromatin, inconspicuous nucleoli, and poorly seen cytoplasm.

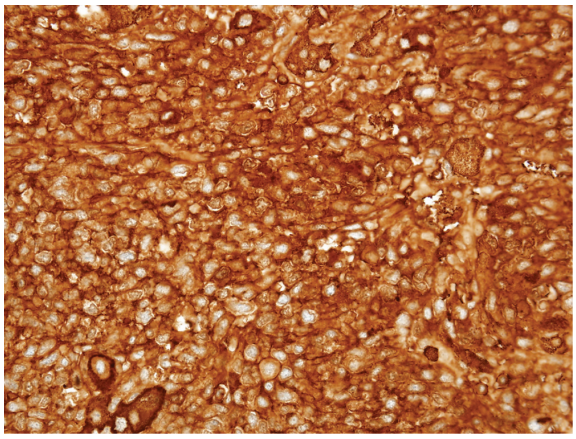

Fig. 4 Chromogranin A immunostain showing diffuse and intense staining.

malignancies[1,2]. Most patients present in the fourth to seventh decades with no sex predilection[1,3-5]. Renal neuroendocrine carcinoma is frequently misdiagnosed with other kidney and urothelial cancer and usually develops in horseshoe kidney[4,5]. The pathogenesis of renal neuroendocrine tumor is still unknown. Several mechanisms have been used to explain the origin of this tumor. It may originate from unrecognized or entrapped neural crest cells in the metanephros during embryogenesis, from neuroendocrine differentiation of primitive totipotential stem cells, pre-existing neuroendocrine cell hypoplasia from metaplastic or teratomatous epithelium, or it arises in association with other congenital renal abnormalities such as horseshoe kidney and polycystic kidney disease[3,4]. The patients commonly present with abdominal pain or mass, flank pain, back pain, hematuria, weight loss, or incidental finding[1,2,4,5]. Renal neuroendocrine tumors rarely secrete hormones or present with paraneoplastic syndrome[2]. However, some patients have evidence of carcinoid syndrome with serotonin-related flushing, edema, and diarrhea[5]. The radiographic findings of renal neuroendocrine tumors hardly distinguish from renal cell carcinoma[4]. They both show solid component, heterogeneity, calcification, and arterial enhancement. Surgery is the treatment of choice for both tumors. The prognosis of renal neuroendocrine tumor depends on stage and tumor differentiation. The common sites of metastasis are lymph nodes, bone, and lung. Five-year survival is 16 to 40% and median survival is 7 to 36 months[2].

The most common cause of ruptured renal tumor is angiomyolipoma, followed by renal cell carcinoma[6]. Three cases have been reported as ruptured neuroendocrine tumors. One case was primary lung cancer and the others were hepatic metastases from transverse colon and maxillary sinus[7-9]. All of them required surgical treatment. To the best of our knowledge, this is the first case report of ruptured malignant neuroendocrine tumor of kidney. Rupture and poor differentiation are the dismal features of neuroendocrine carcinoma. Thus, the disease was rapidly progressed and finally the patient passed away within three months after the operation.

What is already known in this topic?

Renal neuroendocrine tumor is uncommon. The patients may present with abdominal mass, hematuria, paraneoplastic syndrome, or accidental finding. It shows arterial enhancement and calcification on CT scan that is difficult to distinguish from renal cell carcinoma.

What this study adds?

Ruptured renal tumor is usually considered as angiomyolipoma or renal cell carcinoma. This is the first case report of ruptured renal neuroendocrine cancer. Neuroendocrine tumor of kidney is rare. Poorly differentiation and rupture on presentation are dismal parameters.

Potential conflicts of interest

None.

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รายงานผู้ป่วยมะเร็งนิวโรเอนโดครายน์ที่ไตแตก

ชลัยรัชฎ์ สุขอวยชัย, สำเริง รัตนระพี, ธวัชชัย ทวีมั่นคงทรัพย์, ไชยยงค์ นวลยง

ผู้ป่วยหญิงไทย อายุ 51 ปี มาโรงพยาบาลด้วยก้อนที่ท้องด้านขวา ร่วมกับมีอาการปวด และซีด การตรวจทางรังสีพบว่าเนื้องอกของไตด้านขวาแตก ผู้ป่วยได้รับการผ่าตัดไตด้านขวาออก ผลตรวจชิ้นเนื้อพบว่าเป็นมะเร็งชนิดนิวโรเอนโดครายน์ ผู้ป่วยได้รับยาเคมีบำบัด และเสียชีวิตภายหลังการผ่าตัด 3 เดือนต่อมา มะเร็งนิวโรเอนโดครายน์ของไตพบได้น้อยมาก และการแตกของเนื้องอกที่ไตพบได้ไม่บ่อยนัก ส่วนใหญ่พบในมะเร็งของไตและเนื้องอกชนิดแองจิโอมัยโอไลโปมา การแบ่งตัวที่แย่ร่วมกับมีการแตกของเนื้องอกเป็นปัจจัยที่ร้ายแรงของเนื้องอกนิวโรเอนโดครายน์ จากการค้นคว้าพบว่าผู้ป่วยรายนี้เป็นรายแรกที่มีการแตกของมะเร็งชนิดนิวโรเอนโดครายน์ที่ไต