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 angiomylipoma 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

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 angiomylipoma 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
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-contiguous (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. 2](image1.png)
Ruptured tumor at upper pole of right kidney.

![Fig. 3](image2.png)
Hypercellular tumor with hyperchromatic nuclei,
stippled chromatin, inconspicuous nucleoli, and
poorly seen cytoplasm.

![Fig. 4](image3.png)
Chromogranin A immunostain showing diffuse
and intense staining.
malignancies\textsuperscript{(1,2)}. Most patients present in the fourth to seventh decades with no sex predilection\textsuperscript{(1,3-5)}. Renal neuroendocrine carcinoma is frequently misdiagnosed with other kidney and urothelial cancer and usually develops in horseshoe kidney\textsuperscript{(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\textsuperscript{(5,6)}. The patients commonly present with abdominal pain or mass, flank pain, back pain, hematuria, weight loss, or incidental finding\textsuperscript{(1,2,4,5)}. Renal neuroendocrine tumors rarely secrete hormones or present with paraneoplastic syndrome\textsuperscript{(5)}. However, some patients have evidence of carcinoid syndrome with serotonin-related flushing, edema, and diarrhea\textsuperscript{(5)}. The radiographic findings of renal neuroendocrine tumors hardly distinguish from renal cell carcinoma\textsuperscript{(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\textsuperscript{(5)}. The most common cause of ruptured renal tumor is angiomyolipoma, followed by renal cell carcinoma\textsuperscript{(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\textsuperscript{(7,8)}. All of them required surgical treatment. To the best of our knowledge, 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.

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.

References
รายงานผู้ป่วยมะเร็งนิวโรเอนโดครายที่ไตแตก

ชลัยรัชฎ สุขอวยชัย, สำเร็จ รัตนบรรทิพ, ธวัชชัย ทวีมั่นคงทรัพย์, ไชยยงค์นวลยง

ผู้ป่วยหญิงไทย อายุ 51 ปี นั่งรถมาตั้งโต๊ะสูงที่ต้องด้านขวา ร่วมกันมีอาการปวด  gotta การตรวจทางรังสีพบเนื้องอกของไตด้านขวาตก ผู้ป่วยได้รับการผ่าตัดไตด้านขวาออก ผลตรวจชิ้นเนื้อพบว่าเป็นมะเร็งนิวโรเอนโดคราย ผู้ป่วยได้รับยาเคมีบําบัด  และเสียชีวิต 3 เดือนต่อมา มะเร็งนิวโรเอนโดครายของไตได้น้อยลงมาก และการแตกของเนื้องอกที่ไตผัดได้ไม่ต่างกัน ส่วนใหญ่พบในมะเร็งของไตและเนื้องอกนิวโรเอนโดครายโดยไม่ได้ร่วมกัน การแตกเดิ่นที่เกิดรวมกับการแตกของเนื้องอกเป็นปัจจัยที่รวมเจริญของเนื้องอกนิวโรเอนโดคราย จากการค้นคว้าพบว่าผู้ป่วยรายนี้เป็นรายแรกที่มีการแตกของมะเร็งชนิดนิวโรเอนโดครายที่ไต