Case report

CT evaluation of spontaneously ruptured renal angiomyolipomas with massive hemorrhage spreading into multi-retroperitoneal fascia and fascial spaces

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Abstract

Background: Renal angiomyolipomas (RAMLs) can spontaneously rupture and induce hemorrhage that is usually confined to the perirenal space (PS) but may spread beyond the PS into other retroperitoneal fascia and fascial spaces, including up to the subdiaphramatic and down to pelvic extra-peritoneal regions.
Purpose: To evaluate the computed tomography (CT) manifestations of renal angiomyolipoma (RAML) associated with spontaneous rupture and massive hemorrhage spreading beyond the PS into other retroperitoneal fascia and fascial spaces, including up to the subdiaphramatic and down to pelvic extra-peritoneal regions.
Material and Methods: The CT scans of seven patients with spontaneously ruptured of RAMLs and massive hemorrhage (surgically and pathologically confirmed) were retrospectively reviewed. We evaluated the CT signs of the RAML itself and the regions with extensive retroperitoneal spreading after RAML rupture.
Results: The CT manifestations of seven cases with RAML spontaneous rupture and massive hemorrhage included the following: (a) RAML signs: size ($>$4.0 cm, five patients; $<$4.0 cm, two patients), location (periphery, six patients; central portion, one patient), component (fat tissue included, seven patients), and boundary (poorly revealed, seven patients); and (b) signs of extensive retroperitoneal spreading after RAML rupture: involving the PS and extending beyond the PS (seven patients); spread to the pelvic extraperitoneal space (seven patients); attached to the subdiaphragmatic extraperitoneal region (four patients); and extended to the contralateral retroperitoneal spaces (six patients).
Conclusion: CT scans clearly depict both the primary tumor and complicated signs of a spontaneously ruptured RAML with massive hemorrhage, which can affect other fascial planes and retroperitoneal spaces and can extend upward to the subdiaphragmatic region and downward to the pelvic extraperitoneal region or communicate with the contralateral side.

Keywords: Renal angiomyolipoma, rupture, retroperitoneal space, CT

Submitted October 25, 2011; accepted for publication April 30, 2012

Renal angiomyolipoma (RAML), a common benign tumor of the kidney, can spontaneously rupture and induce hemorrhage. The hemorrhage is usually limited to the perirenal space (PS) but, in some cases, may spread beyond the PS and involve the other retroperitoneal fasciae and fascial spaces. Here, we report seven cases of spontaneously ruptured RAML with massive hemorrhage. We emphasized the correlations between the CT manifestations and the anatomical bases of the spreading routes as well as the clinical signs. We hope that our limited experience with seven cases can provide some detailed imaging information for therapeutic planning.

Material and Methods

Imaging and clinical findings in seven patients who had confirmed RAML with rupture and hemorrhage and presented to our institution between 1999 and 2008 were analyzed retrospectively. Five male patients and two female

Table 1 The clinical findings of seven cases of RAML with rupture and hemorrhage

Case	Age (years)	Symptoms	Physical examination signs
1	63	1-week history of left loin pain, with aggravation for 2 days	Left abdominal and lumbar muscle tumefaction and tenderness
2	56	1-day history of right loin pain	Flat belly, with tenderness and tumefaction
3	62	Sudden onset of left loin and flank pain for 6 h	Left abdominal and lumbar muscle tenderness and tumefaction
4	44	Sudden onset of right loin pain for 20 h	Right abdominal and lumbar muscle tenderness and round tenderness; dullness to percussion
5	52	6-h history of acute left upper-quadrant abdominal pain	Left renal swelling and tenderness
6	52	Sudden onset of left loin and flank pain for 9 h	Left abdominal and lumbar muscle tenderness and tumefaction
7	32	2-day history of right loin pain	Right abdominal and lumbar muscle tenderness

patients ranging from 32 to 63 years old were included. This study was approved by our institution's review board and conducted according to its ethical standards. The patients' clinical symptoms and physical examinations and the surgical and pathological evaluation of the primary tumor are shown in Tables 1 and 2.

Four of the seven patients were examined with a 4-channel CT scanner (Somatom Plus-4 Version A, Siemens, Erlangen, Germany), and three patients were examined with a 64-channel multislice CT scanner (Brilliance 64, Philips, Best, The Netherlands). Iodinated contrast material (100–120 mL; Ultravist 300 mgI/mL, Bayer Schering Pharma AG, Berlin, Germany) was administered at a rate of 2.0 to 3.0 mL/s IV in five cases. The CT scan covered the area from the diaphragm to the symphysis pubis in all cases.

All CT images of each patient were reviewed by consensus by two radiologists with 7 and 50 years of experience in abdominal radiology, respectively. The signs of the primary tumor and the routes of spreading in the retroperitoneum after RAML rupture were evaluated.

Results

Clinical presentations

Of the seven patients, three presented with a sudden onset of loin and flank pain, and four patients had suffered from loin and abdominal pain ranging from 6 h to 7 days. The main signs revealed upon physical examination were abdominal and/or lumbar muscle tenderness and tumefaction.

Table 2 Surgical and pathological results for seven cases of RAML with rupture and hemorrhage

Case	Surgical treatment	Pathology results
1	Left nephrectomy and hematoma removal	Left kidney: RAML with hemorrhage
2	Right nephrectomy	Right kidney: RAML
3	Left partial nephrectomy	Lower portion of the left kidney: RAML
4	Right nephrectomy, partial right adrenal gland excision and hematoma removal	Right kidney: RAML with hemorrhage and necrosis Right adrenal gland: not affected by the RAML
5	Left partial nephrectomy	Left kidney: RAML
6	Left nephrectomy and hematoma removal	Left kidney: RAML with hemorrhage
7	Right nephrectomy	Right kidney: RAML with hemorrhage

CT manifestations

The CT manifestations of the tumor itself and the route by which the RAML spontaneous rupture spread throughout the retroperitoneum, with massive hemorrhage in seven cases, are shown in Tables 3 and 4. Five of the seven patients had tumors larger than 4.0 cm; two patients had tumors smaller than 4.0 cm. The tumor boundary in all seven patients was poor. CT images for all patients showed that the tumor sites were located close to the boundary of the kidney, grew near the renal capsule and protruded into the PS. The exception was one lesion located in the central portion of the left kidney and near the renal hilum (Fig. 1).

The CT scans clearly revealed signs of the ruptured tumor associated with hemorrhage, which could easily involve the PS, bridging septa, anterior renal fascia (ARF), posterior renal fascia (PRF), laterocoronary fascia (LCF), posterior pararenal space (PPS), pelvic extraperitoneal space (PES), psoas muscle, and iliac muscle; in addition, anterior pararenal space (APS), subphrenic extraperitoneal region (SER) and contralateral side involvement were also observed in some cases (Table 4). In six cases, the lesions spread into the contralateral retroperitoneal spaces (RS) and across the midline, anterior to the aorta and vena cava; furthermore, the lesions entered the contralateral PPS through a defect in the contralateral PRF in two cases. In four cases, the subcutaneous tissue was involved (Figs. 2 and 3). In four cases, the lesions spread superiorly to the SER, involving the hepatic bare area and/or gastric bare area and the ipsilateral adrenal gland (Fig. 4). In two cases, the hemorrhage broke through the ARF and involved structures in the APS, including the pancreas (Fig. 5).

Discussion

RAML is generally asymptomatic, but some patients may present with mild abdominal pain, palpable masses or hematuria. Patients with ruptured RAMLs often present acute pain as a result of hemorrhage; up to 20% of patients are in shock at the time of the initial presentation (1). Hemorrhages that occur suddenly in association with acute shock can be life-threatening, and their clinical manifestations are easily misunderstood. We strongly suggest

Table 3 CT manifestations of primary signs of RAML with rupture and hemorrhage in seven cases

Case	Mass Size (cm)	Location	Fat component	Shape	Boundary
1	4.0 × 4.3 × 4.0	Anterior and medial portion of the left kidney	Yes	Round	Poor
2	1.9 × 2.0 × 2.2	Medial portion of the upper pole of the right kidney	Yes	Round	Poor
3	3.0 × 3.7 × 4.8	Anterior and lower portion of the left kidney	Yes	Round	Poor
4	10.9 × 16.0 × 16.1	Posterior and upper part of the right kidney	Yes	Irregular	Poor
5	5.8 × 6.8 × 7.2	Posterior and upper part of the left kidney	Yes	Irregular	Poor
6	5.7 × 9.2 × 12.5	Anterior and medial portion of the left kidney	Yes	Irregular	Poor
7	7.2 × 8.1 × 9.0	Anterior and lower portion of the right kidney	Yes	Irregular	Poor

Table 4 CT manifestations for seven cases of RAML rupture and hemorrhage spreading into the RS

Case	PS	Bridging septa	ARF	PRF	LCF	APS	PPS	SER	PES	Psoas muscle	Iliac muscle	Contralateral side
1	+	+	+	+	+	–	+	–	+	+	+	–
2	+	+	+	+	+	–	+	+	+	+	+	+
3	+	+	+	+	+	–	+	–	+	+	+	+
4	+	+	+	+	+	–	+	+	+	+	+	+
5	+	+	+	+	+	+	+	+	+	+	+	+
6	+	+	+	+	+	+	+	+	+	+	+	+
7	+	+	+	+	+	–	+	–	+	+	+	+

APS, anterior pararenal space; ARF, anterior renal fascia; LCF, lateroconal fascia; PES, pelvic extraperitoneal space; PRF, posterior renal fascia; PS, perirenal space; PPS, posterior pararenal space; SER, subdiaphragmatic extraperitoneal region; +, Involved; –, Not involved

that the patients who present at the emergency room with sudden-onset abdominal and loin pain undergo ultrasound or CT/MRI studies.

Angiomyolipomas are composed of abnormal abundant elastin-poor vascular structures, which make these vascular lesions prone to aneurysms and hemorrhage. Generally, the risk of spontaneous perinephric hemorrhage is low when the tumor is <4 cm and increases with increasing tumor size when tumors are >4 cm (2–4). However, two of our cases, tumors <4 cm had ruptured and were associated

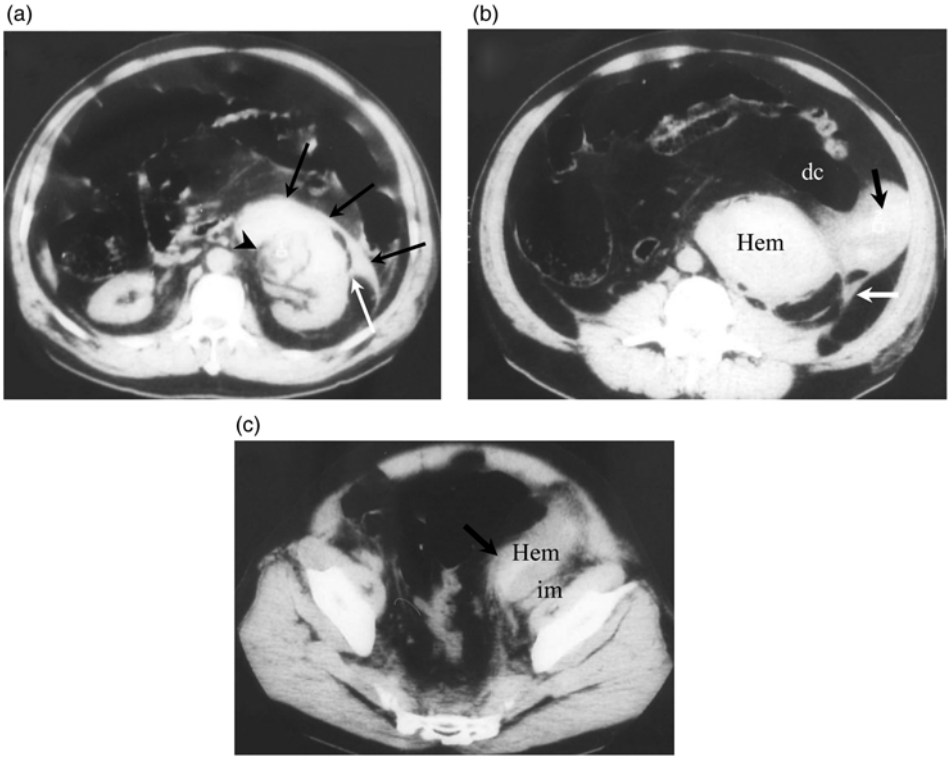

Fig. 1 Case 1: non-enhanced CT scans at the middle kidney (a), infrarenal (b), and iliac fossa (c) levels show a mixed-density mass (4.0 × 4.3 × 4.0 cm) located at the level of the left renal hilum (arrowhead) and associated with a few interior fat components. The tumor had ruptured, and a fresh hemorrhage (Hem) presented high-attenuation (black short arrows) and spread into the left PS, medial to the LCF, posterior to the descending colon (dc), and anteromedial to the iliac muscle (im) into the left PES. The ARF (long, thin black arrow), bridging septa (long white arrow), and PRF (short white arrow) were thickened

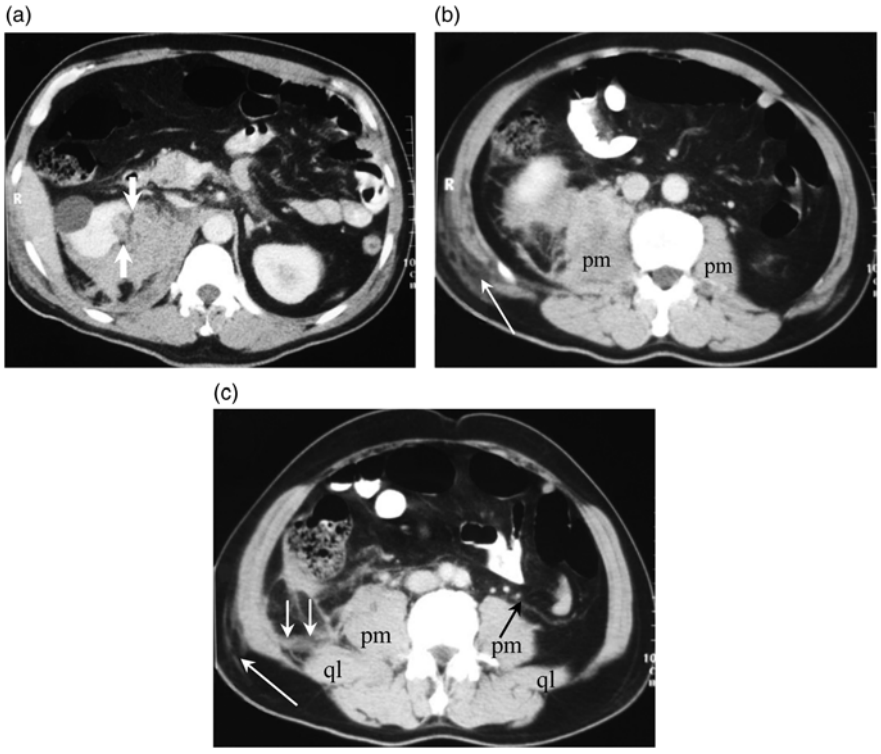

Fig. 2 Case 2: contrast-enhanced CT scans obtained at the upper kidney (a), lower kidney (b), and abdominal aortic bifurcation (c) levels reveals a mixed attenuation nodule (1.9 × 2.0 × 2.2 cm) located at the medial portion of the upper pole of the right kidney (thick white arrows). The tumor ruptured with hemorrhage, extending into the right renal subcapsule and the right PS, abutting the lateral aspect of the psoas muscle (pm), and quadratus lumborum muscle (ql), which appeared swollen. The hemorrhage spread into the right PPS (short white arrows), escaping through the transversalis fascia and the lumbodorsal fascia and resulting in subcutaneous tissue swelling (long and thin white arrows). Thickening of the left ARF is also visible (black arrow)

with obvious hemorrhage. Therefore, small lesions also carry some risk of bleeding (5, 6).

According to the literature, RAML rupture is often limited to within the PS, and hemorrhage involving multiple RSs is rather rare (9%) (5,7). Our seven cases of RAML with spontaneous rupture and hemorrhage showed that hemorrhage can spread extensively beyond the PS into other retroperitoneal compartments and transversely into multiple retroperitoneal fasciae and fascial spaces, such as the ARF, PRF, LCF, APS, and PPS. Such spreading is not only typically ipsilateral but also contralateral (Table 4). Longitudinal spreading may extend as far as the hepatic bare area and/or gastric bare area or down to the iliac fossa, where it becomes life-threatening.

The spreading routes for RAML with hemorrhage are as follows:

1. Extending through the bridging septa: The bridging septa and the perirenal interfascial planes may serve as a bi-directional conduit that allows pathologic processes

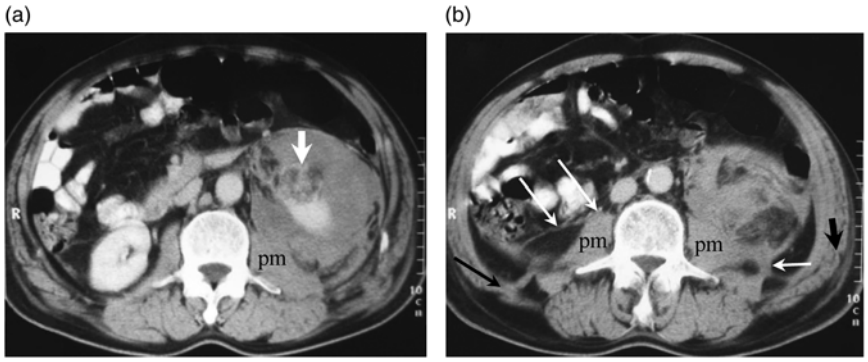

Fig. 3 Case 3: contrast-enhanced CT scans at the lower kidney level (a) and 3.8 cm below (b), show a poorly defined mass (3.0 × 3.7 × 4.8 cm) located at the anteroinferior portion of the left kidney (thick white arrow). An extensive hematoma formed in the left PS and the left renal subcapsule, abutted the lateral aspect of the psoas muscle (pm), spread into the left PPS through the defect in the left PRF (short white arrow), and involved the ipsilateral abdominal wall, resulting in swelling (thick black arrow). Some bleeding crossed the midline anterior to the abdominal aorta and inferior vena cava and extended to the right side (thin white arrows) at the infrarenal level. The right ARF, PRF, and LCF were slightly thickened, and the strip in the right PPS appeared to lie in apposition with the transversalis fascia (thin black arrow)

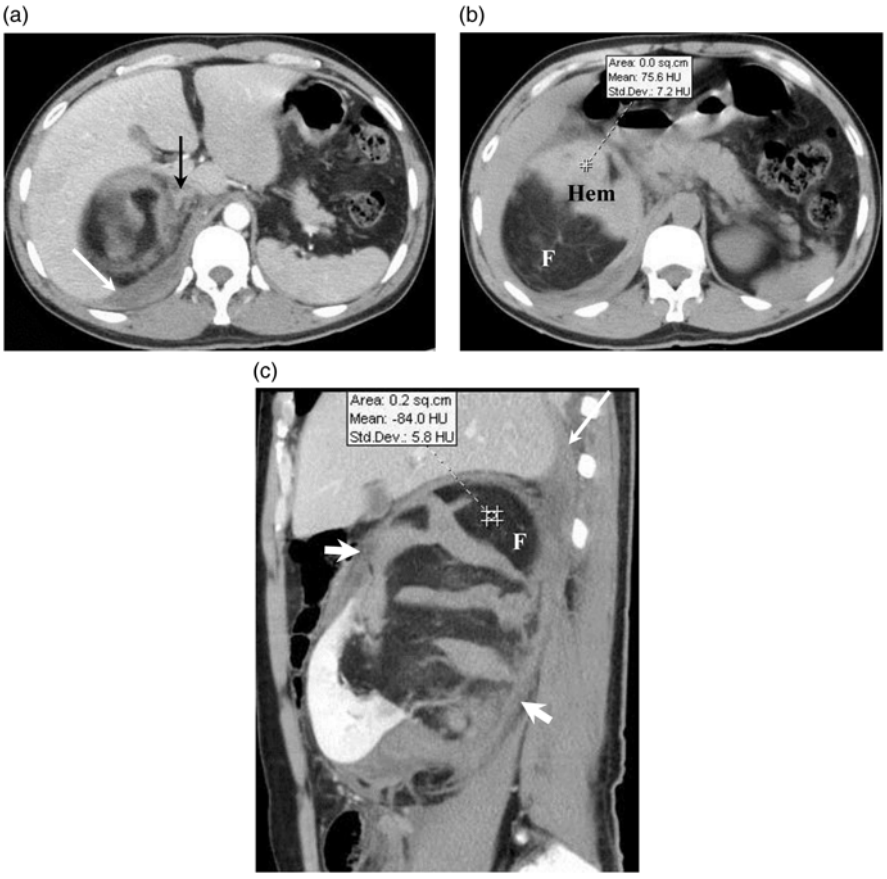

Fig. 4 Case 4: a contrast-enhanced CT scan at the level of the adrenal gland (a), a non-enhanced CT scan of the upper kidney (b), and a contrast-enhanced CT scan sagittal reconstruction through the middle level of the right kidney (c) reveal a very large tumor (10.9 × 16.0 × 16.1 cm) of mixed density, including fat (F), trabecula and hemorrhage, represented by high attenuation (Hem) at the superoposterior part of the right kidney (short, thick white arrows). The tumor projected out of the right kidney and broke into the right renal subcapsule and right PS and then spread upward to the bare area of the liver (white arrow) and the right adrenal gland (black arrow)

to spread from the kidney lesions and the ARF, PRF, and LCF, among other areas (8). Our study demonstrated that bridging septa were involved in all cases and were associated with complete hemorrhage spreading into the laterconal interfascial plane (Case 1).

2. Extending through weak areas of the PRF: Love *et al.* noted that the PRF had weak areas (9). In our seven cases, the CT manifestations of PPS involvement showed that spreading proceeded through weak areas in the PRF. Moreover, some lesions could escape through the lumbar triangle pathway and abdominal muscles into subcutaneous tissue (Figs. 2 and 3), which could explain why the patients felt abdominal and lumbar muscle tumefaction and tenderness, mimicking Grey Turner's sign.

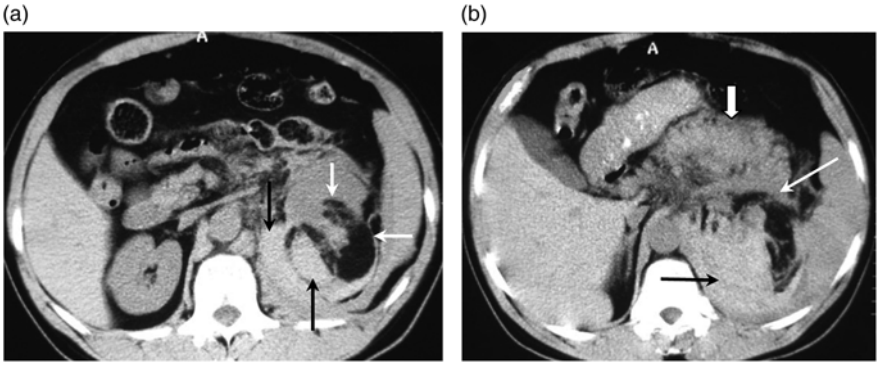

Fig. 5 Case 5: non-enhanced CT scans taken at the middle kidney (a) and pancreatic body (b), levels show a poorly defined mass (5.8 × 6.8 × 7.2 cm) with negative attenuation abutting the adjacent renal parenchyma at the posterior part of the left kidney (short white arrows). The tumor protruded to the left PS. High-attenuation, crescent and irregular bleeding was observed in the posterior portion of the tumor and into the left PS (black arrows). The lesion involved the ARF (long white arrow), and the pancreatic body in the APS (thick white arrow)

Notably, only two cases in our report broke through the ARF and involved APS structures, such as the pancreas (Fig. 5). We presume that the hemorrhage could not readily involve the structures in the APS because of the barrier presented by the ARF, which was relatively solid.

3. Long-distance remote extension: We observed that the hemorrhage lesions spread extensively and even longitudinally to the hepatic bare area and/or gastric bare area (Fig. 4) as well as down to the PES. This spreading could explain the communication among the APS, PPS and PS at the level of the infrarenal space, which in turn connected to the PES (9–11). In the right side, these findings could support the view that the PS had a superior opening that allowed the hemorrhage to extend to the diaphragm and towards the hepatic bare area (12–15). Moreover, the PS may communicate across the midline, anterior to the aorta and vena cava, at the level of the third to fifth lumbar vertebrae (12, 15).

The treatment for a ruptured RAML depends on the tumor size and the severity of hemorrhage. If the tumor is small and limited to the pole of the kidney, it may be removed with enucleation, partial nephrectomy, or anti-shock treatment. However, surgical treatment, including radical nephrectomy and hematoma removal, is the first-line treatment in cases with complete renal involvement and massive hematomas (16). CT scans are particularly valuable in the early recognition of the true extent, complications and subsequent spreading of hemorrhage within the RS and allow surgeons to locate RAMLs that have ruptured and hemorrhaged in the RS. Surgeons can then establish a comprehensive diagnosis and make an appropriate therapeutic management. The ease and safety of CT scanning make it appropriate for follow-up evaluations as well.

In conclusion, CT scans clearly depict the primary tumor signs itself as well as the signs of the ruptured RAML with massive hemorrhage and especially show how the lesions affect the structures in the retroperitoneal fascia and fascial spaces. The correlations between the CT findings and the clinical applications should be investigated further because these correlations are very important for the selection of treatment methods.

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