Malignant peripheral nerve sheath tumour of the renal parenchyma presenting as a fast growing atypical renal cyst

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Abstract

Malignant peripheral nerve sheath tumours (MPNST) of the kidney are very rare, with only 3 cases reported in the English and French literature. However, we report the first case of fast growing atypical renal cyst where a magnetic resonance imaging was an interesting adjunct to the computed tomography scan in imaging this rare tumour.

Introduction

Malignant peripheral nerve sheath tumours (MPNST), also known as malignant schwannoma, arise from Schwann cells which are derived from the neural crest. These tumours are often associated with Von Recklinghausen’s disease. Preoperative diagnosis remains challenging and is based on histology and positive immunohistochemical studies for S-100 protein. Occurrence is usually in the head and neck region or in the limbs. Renal or perirenal MPNST are exceptionally rare findings. We report a case of renal malignant peripheral nerve sheath tumour that highlights the importance of a magnetic resonance imaging (MRI) in such situation.

Case report

A 70-year-old ex-smoking male known for hypertension, diabetes mellitus, dyslipidemia, obesity (body mass index 40) and chronic renal failure (estimated glomerular filtration rate 45.4 mL/min/1.73 m²) was referred to our institution for a growing renal cyst. The patient was asymptomatic and physical examination was unremarkable. Personal and familial history was negative for von Recklinghausen’s disease. Laboratory studies, including complete blood count, electrolytes, urinalysis, glycemia and hepatic profile, were normal except for a high level of serum creatinine and normocytic anemia secondary to chronic renal disease. A computed tomography (CT) scan confirmed multiple simple cysts on both kidneys, except for an 8-cm left mid-upper pole dense cyst. A controlled CT scan 6 months later showed a slight growth and peripheral rim enhancement of this cyst with subtle changes in its content (Fig. 1, part A). An ultrasound confirmed some internal echoes compatible with cellular content. A MRI was recommended by our radiologist 2 months later, and showed a slightly hypointense left renal lesion (Fig. 1, part B) with heterogeneous enhancement following gadolinium administration on T₁-weighted images (Fig. 1, part C). Lesion also appeared heterogeneously hyper-intense on T₂-weighted images. (Fig. 1, part D). Considering the differences between the CT and the MRI images, our radiologist suggested a repeat abdominal CT scan 3 months later which showed significant growth of the left renal mass to 14 cm with heterogeneous internal enhancement (Fig. 1, part E) suggesting a renal tumour. There was focal fat stranding involving the perirenal space. The renal vein remained permeable and without hydronephrosis.

The patient underwent a transperitoneal laparoscopic radical nephrectomy. Lymph nodes were explored during surgery and none were found. Ipsilateral adrenal gland was not removed because it was radiologically normal.

Gross examination of the specimen showed a 17 × 14 × 10-cm kidney weighting about 3 kg. The tumour was gray-white, 15 cm in length and occupied almost all renal parenchyma (Fig. 2). Macroscopically, the tumour infiltrated perirenal fat and was in proximity to the fascia of Gerota without reaching it.

Histological examination (Fig. 3, part A) revealed a sarcoma with fusiform cell. Mitotic count showed multiple zones with >10 mitosis per high field power. Multiple areas of tumour necrosis were found. Adjacent renal parenchyma was normal, except for glomerular hyperfiltration related to chronic renal failure. No vascular emboli were present.
and surgical margins were negative. Immunohistochemical studies of the tumour showed strong S-100 immunoreactivity (Fig. 3, part B) consistent with MPNST and 40% to 50% of tumour cells were positive for Ki-67 (Fig. 3, part C).

Six months after surgery, the CT scan revealed a 7-cm ipsilateral renal fossa tumour recurrence, as well as a metastasis to the ipsilateral adrenal and pancreas. The patient died 10 months after his surgical procedure.

Discussion

MPNST are exceptionally rare tumours with an incidence of 1 to 10 per 1 000 000, representing between 3% and 12% of all soft tissue sarcomas.1,2 These tumours are usually found in patients between 20 to 50 years old, in the head and neck region, trunk or in the limbs.2-5 MPNST show poor prognosis, with reported rates of 5-year disease specific survival of 16% to 60%.1-5 The most effective treatment remains surgical resection and margin status is of primary importance.2-4,6 Conflicting evidence suggests that adjuvant radiotherapy might play a role in the management of MPNST2,3,6 However, no studies found that chemotherapy improves survival.1-3,5,7

We report the fourth case of MPNST in the renal parenchyma (Table 1).8-10 Martinot and colleagues8 reported the first case in 1960 of a 70-year-old male whose left kidney was almost totally replaced by tumour tissue; this patient died on postoperative day 5. Naslund and colleagues9 described an aggressive case of MPNST invading the diaphragm and lung. Surgical margins were positive and the patient declined radiation and chemotherapy. Four months later a 7-cm lesion was found in the renal fossa and the patient eventually died 15 months after surgery. Most recent-
ly, a case of renal MPNST with metastasis to the scalp, lung and shoulder was treated with neoadjuvant doxorubicin and surgical resection of all lesions.10 This patient is free from recurrence at the 2-year follow-up.

Cases of MPNST of the renal capsule were also reported.11-13 Romics and colleagues13 reported a case of MPNST arising from the right renal capsule invading liver and extending up to diaphragm. Surgical margins were positive and the patient developed pulmonary metastasis and died at 3 months following surgical resection. Longer survivals of 2414 and 3615 months were reported in 2 cases of renal pelvis MPNST. Primary16-19 and metastatic20,21 perirenal MPNST have also been reported and tend to present as large renal masses.

Despite the fact that most MPNST are diagnosed in patients with von Recklinghausen’s disease,1-3 no renal or perirenal cases were reported. The diagnosis of renal or perirenal MPNST remains challenging and was established by pathologic examination in all reported cases. As known, the renal CT scan is the gold standard for delineating the

Table 1. Summary of renal and perirenal MPSNT in English and French literature

<table>
<thead>
<tr>
<th>First author</th>
<th>Age/ Sex</th>
<th>Presentation</th>
<th>Size/side, site (cm)</th>
<th>Main treatment</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present case</td>
<td>70/M</td>
<td>Incidental finding</td>
<td>15/left, parenchyma</td>
<td>Laparoscopic radical nephrectomy</td>
<td>Died at 10 mo</td>
</tr>
<tr>
<td>Martinot</td>
<td>70/M</td>
<td>Lower back and flank pain, weight loss, fatigue, fever</td>
<td>NR/left, parenchyma</td>
<td>Radical nephrectomy</td>
<td>Died 5 days postoperative</td>
</tr>
<tr>
<td>Naslund</td>
<td>50/F</td>
<td>Flank pain, weight loss, palpable mass</td>
<td>14/left, parenchyma</td>
<td>Radical nephrectomy</td>
<td>Died at 15 mo with metastasis</td>
</tr>
<tr>
<td>Williams</td>
<td>75/F</td>
<td>Scalp lesion (metastasis)</td>
<td>8/left, parenchyma</td>
<td>Radical nephrectomy</td>
<td>24 mo</td>
</tr>
<tr>
<td>Khandelwal</td>
<td>40/M</td>
<td>Lower back discomfort, palpable mass</td>
<td>NR/lef, renal capsule</td>
<td>Radical nephrectomy</td>
<td>NR</td>
</tr>
<tr>
<td>Jankulovski</td>
<td>65/F</td>
<td>Flank pain, weight loss, palpable mass</td>
<td>13/lef, renal capsule</td>
<td>Radical nephrectomy</td>
<td>6 mo</td>
</tr>
<tr>
<td>Romics</td>
<td>52/M</td>
<td>Flank pain, fever</td>
<td>NR/right, renal capsule</td>
<td>Radical nephrectomy</td>
<td>Died at 3 mo with metastasis</td>
</tr>
<tr>
<td>Voznesensky</td>
<td>46/F</td>
<td>Gross hematuria</td>
<td>4/right, renal pelvis</td>
<td>Laparoscopic nephroureterectomy</td>
<td>36 mo</td>
</tr>
<tr>
<td>Fein</td>
<td>51/F</td>
<td>Flank pain, recurrent pyelonephritis, palpable mass</td>
<td>6/right, renal pelvis</td>
<td>Radical nephrectomy</td>
<td>24 mo</td>
</tr>
<tr>
<td>Pantuck</td>
<td>50/F</td>
<td>Palpable mass</td>
<td>28/right, perirenal</td>
<td>Radical nephrectomy</td>
<td>Died at 42 mo</td>
</tr>
<tr>
<td>Parfitt</td>
<td>63/M</td>
<td>Weakness, fatigue, weight loss, palpable mass</td>
<td>11 and 6/left, perirenal</td>
<td>Radical nephrectomy</td>
<td>9 mo</td>
</tr>
<tr>
<td>Bair</td>
<td>56/M</td>
<td>Microscopic hematuria</td>
<td>7/right, perirenal</td>
<td>Radical nephrectomy</td>
<td>5 mo</td>
</tr>
<tr>
<td>Deming</td>
<td>33/F</td>
<td>Abdominal pain, palpable mass</td>
<td>NR/lef, perirenal</td>
<td>Tumour excision without nephrectomy</td>
<td>Died at 11 mo with metastasis</td>
</tr>
<tr>
<td>Cachay</td>
<td>74/F</td>
<td>Follow-up for malignant subcutaneous schwannoma</td>
<td>9/right, metastasis</td>
<td>Radical nephrectomy</td>
<td>5 mo</td>
</tr>
<tr>
<td>Garcia</td>
<td>73/F</td>
<td>Follow-up for malignant soft-tissue MPNST</td>
<td>8/right, metastasis</td>
<td>Radical nephrectomy</td>
<td>Died at 5 mo</td>
</tr>
</tbody>
</table>

MPNST: malignant peripheral nerve sheath tumours; mo: months, NR: not reported.
nature of a renal mass and MRI remains an alternate modality. However, some MRI findings were reported to be suggestive of benign peripheral nerve sheath tumour (PNST). They included isointensity on T1-weighted images, a high signal intensity on T2-weighted images and strong and homogeneous gadolinium-enhancement on T1-weighted images. We report, however, the first case of malignant tumor with MRI. Although our case had heterogeneous enhancement following gadolinium administration, it shared some features of PNST, such as enhancing outer rim and T2-weighted images that should raise suspicion for MPNST. Finally, the rapid evolution of the radiological features reported in this case and the poor survival shows the aggressiveness of MPNST.

In this case, the discrepancies between CT scan and MRI images might be due to the superiority of MRI for detecting such tumours or to actual tumour growth as images were taken 2 months apart. However, it shows that MRI might help in the differential diagnosis of renal cyst when ultrasonography and CT scan cannot provide a definitive diagnosis. It also highlights the importance of MRI and serial radiological follow-up for atypical renal cyst with rapid growth that might carry poor prognosis and should prompt surgical management.

Conclusion

We reported a rare case of MPNST presenting as an atypical renal cyst where MRI provided important information along with the CT scan.

Acknowledgements: We would like to thank Tania Fayad, PhD, for the critical reading of this article.

Competing interests: None declared.

This paper has been peer-reviewed.

References


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