A 72-year-old white female presented with 3-week history of increasing abdominal pain, bilateral leg swelling, decreasing urine output and shortness of breath. She started to have worsening bilateral leg edema right greater than left. She had Doppler ultrasound of her legs which was negative for deep vein thrombosis. She was prescribed furosemide two times daily with minimal improvement in swelling. A few weeks later, she started to have abdominal pain and shortness of breath. Abdominal pain was new onset, dull, right-sided with no radiation or aggravating relieving factors. She noticed abdominal mass incidentally while taking shower which became more apparent on bending. Her past medical history was significant for hypertension, coronary artery disease with bare-metal stent to the right middle coronary artery 9 years ago and insulin-dependent diabetes mellitus.

On arrival, her vitals were 36.4°C, 100/68 mm Hg, 97 per min, 31 per min respiratory rate and body mass index 46 kg/m². Abdominal examination was notable for a right-sided antero-lateral mass with irregular borders crossing the midline anteriorly obscuring the lower liver edge. Shifting dullness and fluid thrill were absent. Lower limbs examination was significant for left below knee amputation with significant pitting edema greater on right than left leg. Computed tomography (CT) scan of the abdomen and pelvis showed exophytic predominantly hypodense multiseptated mass from the interpolar region and lower pole of the kidney (Figure 1).

Given the location and size of the mass, the patient underwent right-sided complete nephrectomy with removal of ipsilateral adrenal gland and a single enlarged lymph node. The mass extended to the peri-renal fascia and into the renal pelvic-caliceal system abutting the renal sinus, measuring 31.2 × 24.6 × 14.6 cm and tan-white to orange in color with >70% areas of hemorrhage and necrosis (Figure 2). Histology was suggestive of a high-grade malignant peripheral nerve sheath tumors (MPNSTs) (Figure 2).

MPNSTs are a malignant transformation of Schwann cells arising in the peripheral nerves which have not been known to arise from a pre-existing Schwannoma. They are more usually arising from distal ends of the nerves and less likely to be involving the proximal part. This explains the early diagnosis of distal MPNSTs and significantly better outcomes.1–3 Retroperitoneal Schwannomas and MPNSTs are extremely rare tumors with the overall incidence of MPNST being 0.001% in general population and 8–13% in people with neurofibromatosis type 1 (NF-1).4 In most
cases, these tumors are linked to main lumbar or sacral plexus. However, in majority of cases the associated nerve is hard to identify.

MPNSTs are highly cellular tumors comprised of spindle cells which are reminiscent of Schwann cells. Sometimes it may show variations with rhabdomyosarcomatous differentiation which is very malignant containing embryonic striated muscle component. These aggressive tumors usually occur in association with NF-1, however, sporadic cases are not very uncommon. Those associated with NF-1 and also in sporadic cases, NF-1 homozygous mutation is noted, however, additional mutations are necessary (e.g. p53 and CDKN2A deletions). Interestingly, intense reactivity to p53 has been associated with malignancy. MPNST present early when in association with NF-1 usually in third or fourth decade than in sporadic cases which presents mostly in seventh decade. Incidental masses are the common presentation for these tumors and usually associated with pain and peripheral nerve deficits (i.e. radiculopathy).

The management of these tumors includes wide resection and follow-up to rule out local recurrences. Metastasis noted to involve lymph nodes, liver, brain, bone and adrenals in 20%. These are mostly managed by resection or debulking with clear margins and regular follow-up for recurrence. In recent times, adjuvant radiation therapy has been proposed but effectiveness is still under question as data are limited. There is no established trials to support preoperative or postoperative adjunctive radiation therapy as the reported cases are few. Prognosis is usually poor. Indicators for adverse outcomes are late onset, size >5–7 cm, high-grade tumor differentiation, NF-1 association and lack of tumor free margins post-resection. In one study of 36 patients, 61% lived up to 25 months. Early death was associated with mass size of >7 cm. In another study of 53 patients, 23 had mean survival of 54 months, tumor recurrence has been reported to be 26% within 1–10 years. In our case, the patient tolerated the procedure very well and was discharged home to start adjuvant radiation therapy as outpatient.

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References


