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When the nerves turn malignant: A case of retroperitoneal peripheral nerve sheath tumor: A case report

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Abstract
Introduction: Retroperitoneal malignant peripheral nerve sheath tumors (MPNST) are uncommon soft tissue sarcomas, usually presenting late because of the wide potential space. Preoperative diagnosis is difficult, and treatment mainly consists of surgical resection.
Case Presentation: A 59-year-old man presented with abdominal pain for one month. Contrast-enhanced CT abdomen disclosed a huge heterogeneously enhancing retroperitoneal mass (11.9 × 11.1 × 15.7 cm) in the right hypochondriac and lumbar region, which was pushing the IVC and right renal vein and showing associated lymphadenopathy. The patient had surgical resection of a well-defined encapsulated retroperitoneal tumor with dimensions of 16 × 12 × 9 cm with an attached cord-like structure of about ~10 cm. Histopathology revealed a spindle cell neoplasm with pleomorphism, necrosis, and mitoses (>5/10 HPF). Immunohistochemistry was positive for S-100, Sox10, and HMB45 with loss of SMA/Desmin/CD34. Diagnosis: Low-grade Malignant Peripheral Nerve Sheath Tumor (MPNST).
Discussion: Retroperitoneal MPNSTs are extremely uncommon (<5% of soft tissue sarcomas). Imaging may raise suspicion of sarcoma, yet only histopathology and IHC can make a definitive diagnosis. Complete resection is still the mainstay of treatment. Prognosis is variable and depends on the size of the tumor, grade, extent of resection, and NF1 association.
Conclusion: The diagnostic difficulty of retroperitoneal MPNST is brought out in this case and the need for a multidisciplinary evaluation involving imaging, histopathology, and immunohistochemistry.

Keywords: Retroperitoneal tumor, malignant peripheral nerve sheath tumor, sarcoma, case report

Introduction
Retroperitoneal soft tissue sarcomas account for less than <0.2% of all malignancies, with the most frequent subtypes being liposarcomas and leiomyosarcomas (Strauss 2011) [2]. Malignant Peripheral Nerve Sheath Tumors (MPNSTs) are infrequently found in the retroperitoneum and tend to manifest late after silent growth. They tend to compress major structures such as the IVC, renal vessels, and ureters, causing nonspecific symptoms. Early diagnosis is essential because prognosis is based on early radical resection (Gronchi 2013) [3].

Case Presentation
Patient: 59-year-old man
Symptoms: Dull aching pain in abdomen for 1 month, no jaundice/hematuria.
Examination: Abdominal fullness with firm mass in right hypochondriac & lumbar area.

Investigations
CT Abdomen & Pelvis: Large well-defined heterogeneously enhancing mass (11.9 × 11.1 × 15.7 cm) compressing IVC and right renal vein; multiple necrotic lymph nodes; mild ascites (Fig. 1).

Surgery: Right Makuchi's Incision, removal of encapsulated well-circumscribed retroperitoneal tumor (16 × 12 × 9 cm) with adherent cord-like structure (~10 cm). Nearby structures displaced but not invaded. (Fig 2)

Histopathology (HPE): Malignant spindle cell neoplasm, fascicular growth pattern, hyperchromatic nuclei, mitotic figures >5/10 HPF, necrosis, mucinous degeneration.

Immunohistochemistry (IHC): Positive for S-100, Sox10, HMB45; negative for SMA, Desmin, CD34. Diagnosis: Low-grade MPNST.

Outcome: Patient recovered well postoperatively. On follow-up, no recurrence at 3 months.

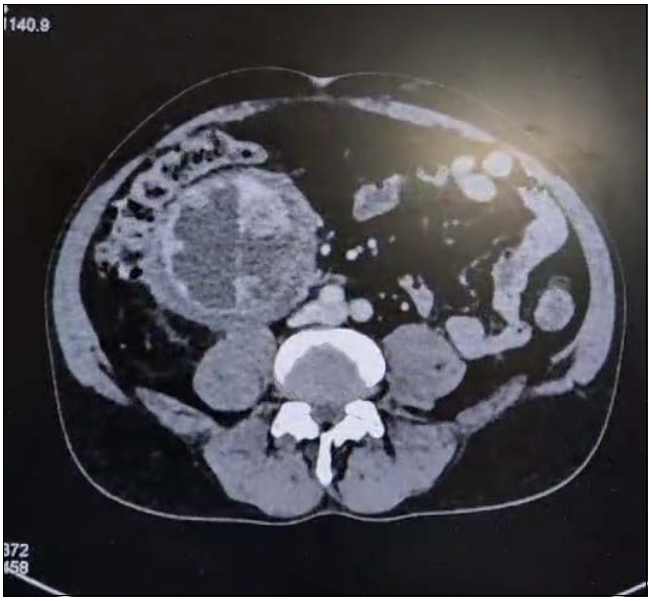
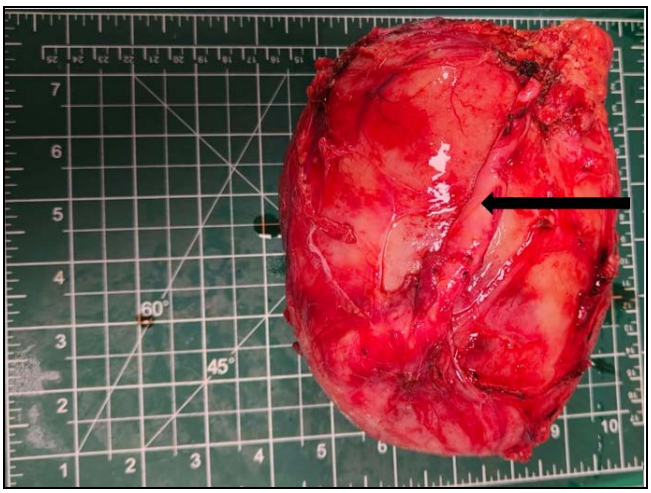


Fig 1: CECT (A+P) Images



Marked Arrow: Nerve

Fig 2: Post-Operative Specimen Photo

Discussion

Rarity: MPNST accounts for ~5–10% of all soft tissue sarcomas; retroperitoneal location is extremely rare (Gatta 2011) [1].

Diagnosis: Imaging (CT/MRI) helps define extent, but cannot differentiate subtypes.

Histopathology with IHC is mandatory. S-100 and Sox10 positivity with spindle morphology supports MPNST (WHO Soft Tissue Tumor Classification, 2020) [4].

Management

Negative margin complete surgical excision is gold standard. Adjuvant radiotherapy/chemotherapy role is controversial but is taken in high-grade or unresectable tumors.

Prognosis: Bad, with 5-year survival ~34–50%; tumor size >5 cm, high grade, necrosis, incomplete resection, and association with NF1 are prognostic factors.

Conclusion

Retroperitoneal MPNSTs are uncommon tumors, which create diagnostic and therapeutic problems. Imaging is suggestive of sarcoma, but histopathology and IHC are absolute. Complete surgical excision gives the best result.

Informed Consent

Written informed consent was sought from the patient for publication of this case and images.

Highlights

- Retroperitoneal malignant peripheral nerve sheath tumors (MPNSTs) are extremely rare, accounting for <0.2% of all malignancies.
- These tumors often present late due to the silent expansion capacity of the retroperitoneal space.
- Imaging (CT/MRI) defines tumor extent and vessel involvement, but definitive diagnosis requires histopathology and immunohistochemistry.
- Complete surgical excision with negative margins remains the cornerstone of treatment, even in large tumors compressing major vessels.
- This case highlights the diagnostic challenge and surgical management of a giant retroperitoneal MPNST mimicking an abdominal mass.

Conflict of Interest

Not available

Financial Support

Not available

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