

Retroperitoneal Hybrid Nerve Sheath Tumor Mimicking Malignant Imaging Manifestations: A Rare Case Report

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Case Report

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Abstract

Background: Hybrid peripheral nerve sheath tumors (hPNSTs) is an exceedingly rare benign peripheral nerve sheath tumor, featuring elements from at least two nerve sheath tumor cells. We herein report a rare case of retroperitoneal HNST manifesting malignant imaging features.

Case presentation: We reported a 40-year-old female presented with unexplained abdominal pain, with no significant personal or familial medical history. Imaging workup revealed a large solid-cystic retroperitoneal mass with internal hemorrhage, calcification and cystic degeneration. The lesion showed restricted diffusion and progressive delayed enhancement on MRI, with blurred borders adjacent to major retroperitoneal vessels and the right ureter, highly suggestive of an invasive malignant tumor. The patient underwent complete surgical resection. Postoperative histopathology and immunohistochemistry confirmed a benign hybrid schwannoma/neurofibroma, with no lymph node metastasis identified. The patient recovered uneventfully and remained recurrence-free during follow-up.

Conclusions: Retroperitoneal hPNST can exhibit atypical imaging features that closely resemble malignant neurogenic tumors. Accurate diagnosis relies on the combined analysis of radiological findings and immunohistochemical results. This case expands the known anatomical and imaging spectrum of hPNSTs, improves clinicians' overall recognition of this rare disease.

Background

Hybrid peripheral nerve sheath tumors (hPNSTs) were formally categorized in the 2016 WHO Classification of Tumours of the Central Nervous System[1], represented a distinct category of benign peripheral nerve sheath neoplasm. To date, fewer than 50 cases have been documented globally[2]. These lesions are defined by composite morphological features consisting of at least two types of nerve sheath tumors, including neurofibroma-perineurioma, schwannoma-perineurioma, as well as neurofibroma-schwannoma[3]. Among these subtypes, hybrid schwannoma/perineurioma is the most prevalent, followed by hybrid schwannoma/neurofibroma, whereas hybrid neurofibroma/perineurioma is relatively uncommon. Reported cases of hPNSTs predominantly arise in the extremities and extracranial branches of cranial nerves[4]. Clinically, these tumors pursue an indolent clinical course with a low recurrence rate after complete surgical resection, and malignant transformation is extremely rare[3]. Owing to the scarcity of clinical cases, the imaging characteristics of hPNSTs remain poorly characterized. We herein report an extremely rare case of hPNST presenting prominent malignant imaging manifestation.

Case presentation

Clinical presentation

A 40-year-old female patient presented with abdominal pain, with no identifiable predisposing cause, no remarkable past medical history or positive family history. Physical and neurological examinations

revealed no abnormal findings. All baseline hematological and urinary laboratory findings were within the normal reference range.

Abdominal CT (Fig. 1) identified a round, slightly hypodense mass in the right retroperitoneum, measuring approximately 10.6 cm in maximum diameter. Cystic, hemorrhage, and calcifications were observed within the lesion. The mass showed heterogeneous enhancement and contained thick blood vessels after contrast administration. The lesion showed indistinct demarcation from the right ureter. The right kidney presented with hydronephrosis and decreased enhancement. Besides, the right kidney was encased by cystic lesions containing internal septa. Subsequent MRI (Fig. 2) further characterized the retroperitoneal lesion. The lesion showed hypointense signals on T1-weighted imaging (T1WI) and hyperintense signals on T2-weighted imaging (T2WI). The solid components of this tumor exhibited high signal intensity on DWI with corresponding decreased ADC values, and demonstrated “delay enhancement” strengthened on contrast enhancement. The inferior vena cava, right renal vein and right ureter were poorly delineated. In addition, a crescent-shaped lesion around the right kidney exhibiting hyperintense signals on T1WI and T2WI and peripheral enhancement. Based on these aggressive imaging manifestations, a preoperative diagnosis of invasive malignant retroperitoneal and perirenal tumor was highly suspected.

No other systemic tumors were detected during hospitalization. Percutaneous biopsy confirmed a neurogenic tumor, highly suggestive of schwannoma. Immunohistochemical results were as follows: S-100 (+), SOX10 (+), H3K27me3 (+), Ki-67 (1% positive), and HMB45 (-). The patient subsequently underwent retroperitoneal tumor resection. Intracapsular tumorectomy was performed according to the preoperative pathological suggestion.

Postoperative histopathology reconfirmed a retroperitoneal neurogenic tumor with solid-cystic components, and coagulum-like substances were found in the cystic areas. Combined with clinical data and immunohistochemical profiles, the final diagnosis was hybrid schwannoma/neurofibroma. Immunohistochemical staining showed diffuse positivity for S-100 and SOX10, focal H3K27me3 positivity, and 2% Ki-67 proliferative activity. Markers including SMA, EMA, Desmin, Pan-cytokeratin, CD34 and Synaptophysin were all negative. Three resected regional lymph nodes were examined, and no metastatic tumor cells were found (0/3) (Fig. 3).

The patient achieved a favorable recovery during the follow-up CT examinations after surgery, without any residual neurological deficits.

Discussion and conclusions

hPNSTs are rare benign nerve sheath tumors composed of dual or multiple differentiated components. Given the extremely limited number of reported cases, no typical or standardized imaging features have been established for this entity. The present case exhibited even more atypical imaging manifestations, representing a rare presentation of this uncommon tumor.

Histologically, hPNSTs combine different nerve sheath tumor components. Schwannomatous areas feature ordered spindle cells, Verocay bodies and hyalinized vessels, while neurofibromatous areas show wavy nuclei and collagen-rich myxoid stroma[5]. Perineuriomatous components typically show fascicular or onion-bulb layered architecture with stromal collagenization. This mixed histology overlaps substantially with conventional benign nerve sheath tumors, making immunohistochemistry essential for accurate subtyping. Schwannomatous components show diffuse strong S-100 and SOX10 expression, while neurofibromatous areas contain a mixed cell population of S-100/SOX10-positive tumor cells and CD34-positive stromal fibroblasts. EMA, claudin-1 and GLUT-1 positivity indicates perineurial differentiation, and focal SMA expression can be detected in sclerotic perineurioma[6]. Consistent with these published findings, our case showed diffuse S-100 and SOX10 immunoreactivity, focal H3K27me3 staining and a low Ki-67 index, whereas mesenchymal and epithelial markers were all negative. These results confirmed the composite schwannoma/neurofibroma components and benign biological nature of the lesion.

Previous studies have demonstrated that most hPNSTs arise in the soft tissues of the extremities[7] and trunk[8]. With accumulating case reports, hPNSTs have also been identified at rare anatomical sites, including subcutaneous tissue[9], gastrointestinal tract[10] and orbit[11, 12]. Hayashi et al.[13] described an intraspinal hybrid schwannoma/perineurioma with high proliferative activity, obvious mitotic figures and a Ki-67 index of 20%, which was associated with postoperative recurrence. Blake et al.[14] reported 11 cases of hybrid schwannoma/neurofibroma in patients with neurofibromatosis type 2. Among these reported cases, schwannoma/perineurioma is the most common subtype, followed by schwannoma/neurofibroma.

The limited number of previously published imaging reports on hPNSTs have described marked contrast enhancement with intralesional cystic change[3, 4, 11, 12]. This tumor presented intratumoral hemorrhage, calcification and cystic degeneration, without a complete capsule, target sign or nerve entry/exit sign. Solid tumor components showed restricted diffusion and delayed contrast enhancement. Notably, the tumor lacked clear demarcation from the adjacent inferior vena cava and right ureter, mimicking invasive malignant growth, which easily led to false preoperative malignant diagnosis.

Classic schwannomas typically demonstrate marked homogeneous or heterogeneous enhancement after contrast injection. In contrast, this our hPNST showed only mild-to-moderate enhancement, which may be attributed to abundant fibrous and collagenous stroma within the lesion. Conventional schwannomas possess well-organized Antoni A/B zonation, perivascular hyalinization and an intact EMA-positive capsule[15]. However, hPNSTs lack canonical Antoni A/B layered architecture and complete tumoral capsule, with heterogeneously distributed mixed cellular components, ultimately resulting in atypical imaging phenotypes[5, 16]. In contrast, Malignant peripheral nerve sheath tumors (MPNSTs) usually present as large, irregular masses with more heterogeneous signal intensity, frequently accompanied by peritumoral edema and invasive margins[17]. The substantial imaging overlap between this hPNST and MPNST creates considerable challenges for preoperative differential diagnosis. DWI and quantitative ADC values provide valuable auxiliary evidence for distinguish. MPNSTs

exhibit severe diffusion restriction due to high cellular density, with ADC values generally below 1.0×10^{-3} mm²/s[18]. The ADC value of the current tumor was higher than this threshold.

Conclusions

In conclusion, the actual incidence of hPNSTs is likely underestimated in clinical practice, mainly due to its rarity and atypical imaging manifestations. This case supplements the limited imaging data of retroperitoneal hPNSTs and confirms that benign hPNSTs can closely mimic malignant neurogenic tumors on preoperative imaging. Our findings highlight a noteworthy clinical diagnostic pitfall, further accumulation of clinical, radiological and pathological data.

Abbreviations

hPNSTs Hybrid peripheral nerve sheath tumors

CT Computed tomography

MRI Magnetic resonance imaging;

MPNSTs Malignant peripheral nerve sheath tumors.

Declarations

Ethics approval and consent to participate

The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). Patient presented in this report was investigated within a retrospective study approved by the ethical review board of Zhongshan Hospital (Xiamen branch), University of Fudan University (Protocol ID Number B2025-317R).

Consent for publication

Written informed consent was obtained from the patient's legal guardian for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing interests

The authors declare that they have no competing interests.

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Author Contribution

LZ performed the background literature review, prepared figures 1-2, drafted the manuscript and performed manuscript revisions. LR performed histopathological specimen analysis, prepared figures 3, presentation and manuscript preparation. All authors reviewed the manuscript.

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N/A.

Data availability

Supporting data are available from the corresponding author upon reasonable request.

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Figures

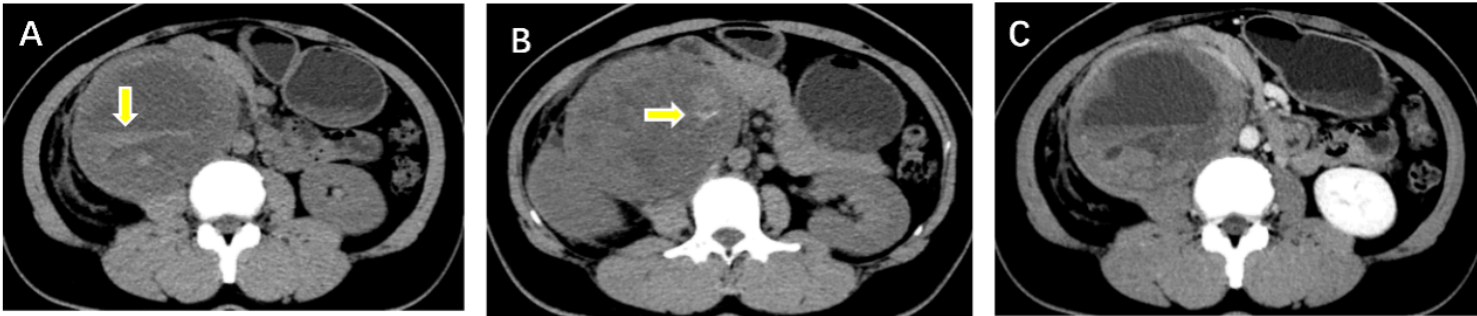


Figure 1

Contrast-enhanced computed tomography (CT). (A) Intratumoral haemorrhage (arrow); (B) intratumoral calcification (arrow); (C) heterogeneous mild enhancement of the lesion.

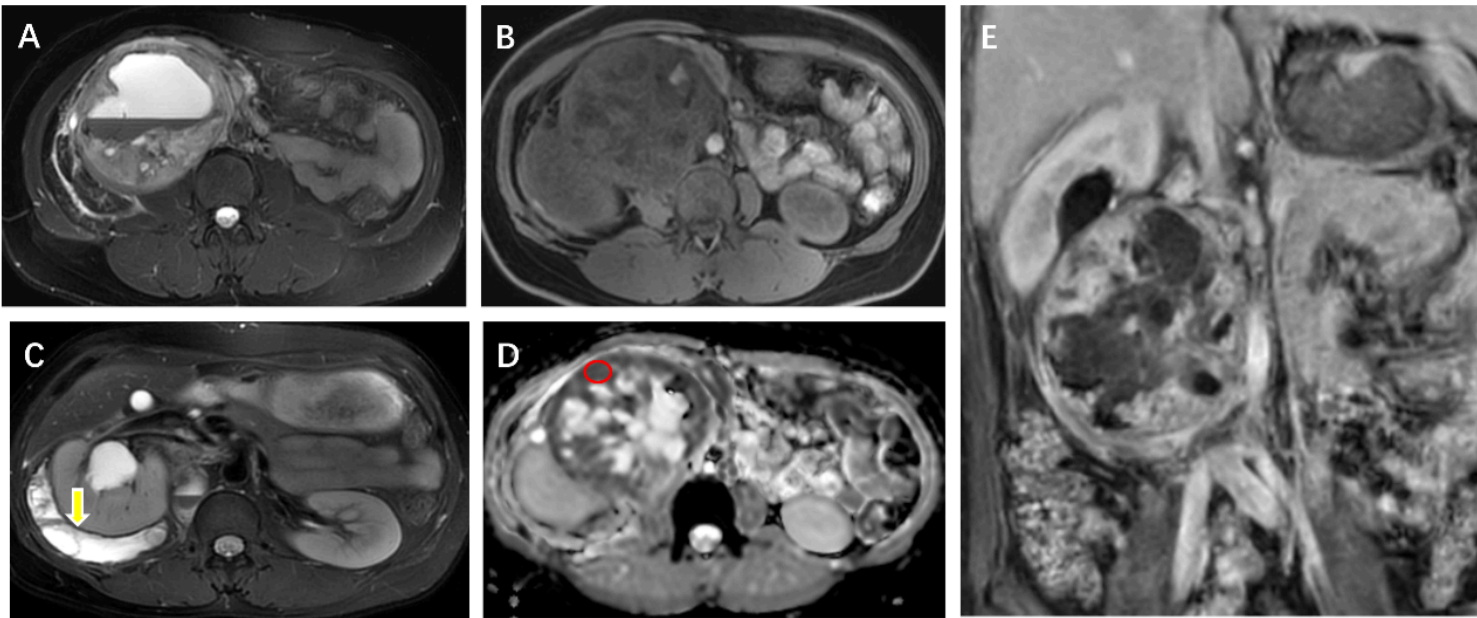


Figure 2

Magnetic resonance imaging (MRI). (A, B) T2WI and T1WI sequences show heterogeneous signal intensity due to intratumoral haemorrhage. (C) T2WI demonstrates tumour encasement of the perinephric space surrounding the right kidney (arrow). (D) ADC map: the solid component (circle) shows an ADC value of approximately $(1.57 \pm 0.11) \times 10^{-3} \text{ mm}^2/\text{s}$. (E) Contrast-enhanced imaging reveals ill-defined borders between the lesion and the inferior vena cava, right renal vein and right ureter.

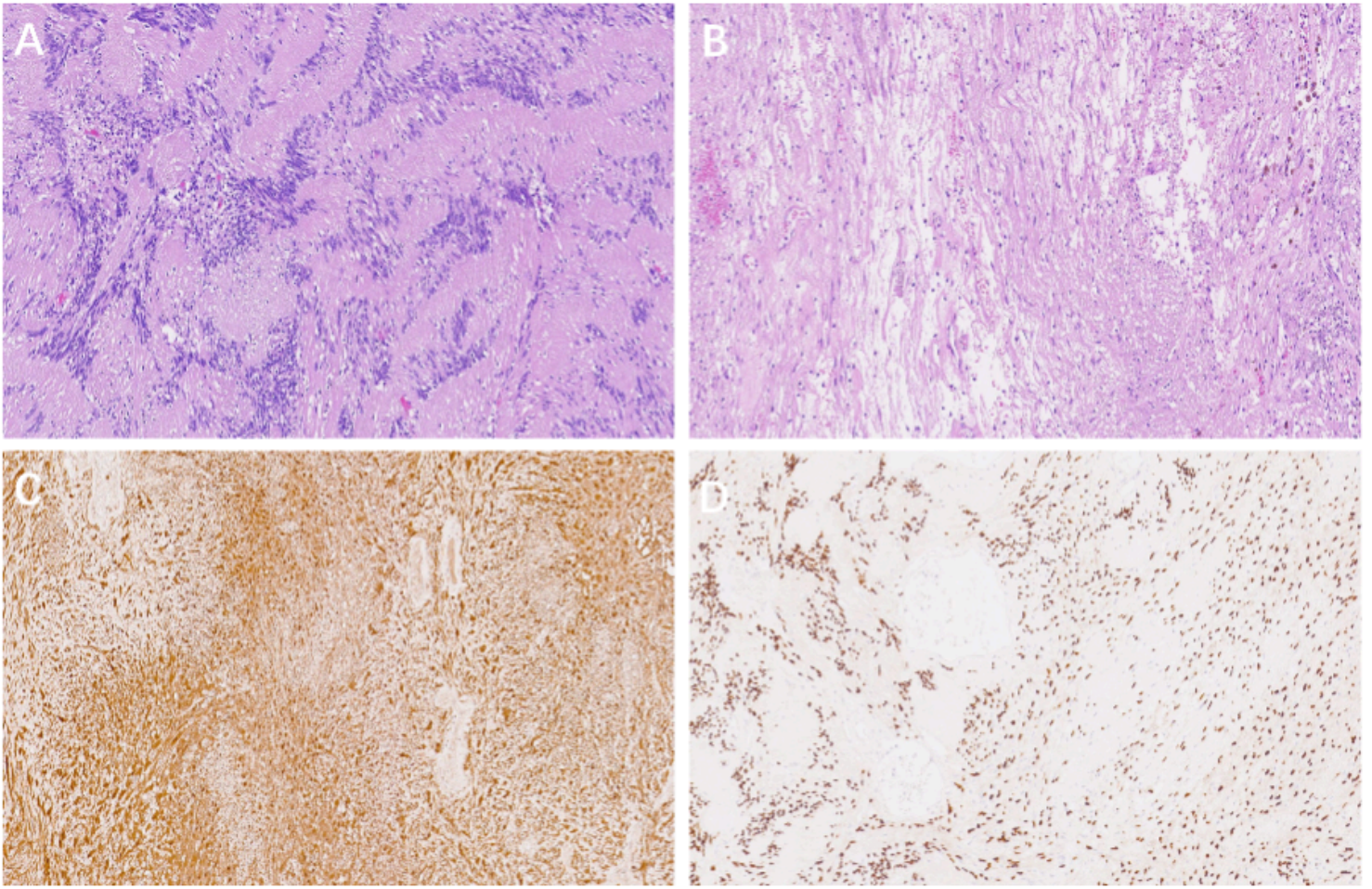


Figure 3

(A) Schwannomatous regions contain orderly arranged spindle cells and hyalinized blood vessels. (B) Neurofibromatous regions exhibit wavy nuclei within collagen-rich myxoid stroma. (C, D) Immunohistochemical staining demonstrates tumour cells positive for S-100 and SOX10. All photomicrographs, $\times 100$ magnification.

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