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CASE REPORT

CANCER INNOVATION

A case report of multimodal ultrasound imaging in the diagnosis of giant retroperitoneal ganglioneuroma

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Abstract

Retroperitoneal ganglioneuroma is a rare benign tumor that is challenging in terms of clinical diagnosis. Computed tomography and magnetic resonance imaging are usually performed for diagnosis rather than convenient and inexpensive ultrasonography. Here, we present the case of a 21-year-old female patient who was diagnosed by multimodal ultrasound imaging and whose diagnosis was confirmed by ultrasound-guided core needle biopsy before surgery. We hope that this rare case will help clinicians and radiologists realize the advantages of multimodal ultrasound imaging in the diagnosis of retropeitoneal solid tumors, and reduce misdiagnosis.

KEYWORDS

diagnosis, ganglioneuroma, retroperitoneal mass, ultrasonography

1 | INTRODUCTION

Retroperitoneal ganglioneuroma (GN) is a benign tumor originating in the sympathetic nervous system, accounting for only 0.72%–1.6% of primary retroperitoneal tumors [1]. It is difficult to diagnose due to its nonspecific clinical presentation [2]. Unlike retroperitoneal sarcoma, GN have a good prognosis and low rates of recurrence and malignancy. Therefore, proper diagnosis is of great value for proper management. However, such tumors have been mainly reported on computed tomography (CT) and magnetic resonance imaging (MRI) examinations, and the use of ultrasonography is rarely reported. In this study, a giant GN diagnosed by multimodal ultrasound imaging is reported to improve the understanding of rare tumors, broaden the idea of differential diagnosis of retroperitoneal solid tumors in ultrasonography, and reduce misdiagnosis.

2 | CASE PRESENTATION

The patient, a 21-year-old woman, was diagnosed with a giant tumor behind the right peritoneum by CT scan within 5 days. The patient did not have fever, abdominal pain, bloating, and weight loss. Physical examination revealed a palpable mass in the right upper quadrant without tenderness. Important tumor markers, such as α-fetoprotein and carcinoembryonic antigen, were normal. B-mode ultrasonography (US) showed a 19.2 cm × 10.7 cm hypoechoic mass in the hepatorenal space with clear margins and multiple punctate calcifications inside. When

Abbreviations: CEUS, contrast-enhanced ultrasonography; CT, computed tomography; GN, ganglioneuroma; MRI, magnetic resonance imaging; US, ultrasonography.

the patient was asked to breathe, the mass was found to show asynchronous movements along with the liver and the right kidney. Color Doppler showed only a small amount of blood flow signals within the lesion. Conventional ultrasonography suggested a homogeneous retroperitoneal mass with calcifications, and contrast-enhanced ultrasonography (CEUS) was recommended. After an injection of 2.4 mL of contrast agent SonoVue™ (Bracco), the lesion began to enhance at the 6th second, followed by a mild enhancement pattern, significantly weaker than the right kidney, and persistent enhancement after 2 min (Figure 1). CEUS indicated a benign lesion and neurogenic neoplasms were not excluded. Contrast-enhanced CT showed a hypodense mass with well-defined margins and microcalcifications in the retroperitoneal space. Uneven enhancement of flocculent was shown in the venous phase. MRI showed a low T1 signal, a heterogeneous high T2 signal, and medium-high DWI signal intensity, with a gradual and mild enhancement of delayed images (Figure 2). It replaced the inferior vena cava, right kidney, and liver without definite invasion. Differential diagnoses included retroperitoneal sarcoma, neurogenic tumors, and teratoma. Ultrasound-guided core needle biopsy with CEUS revealed mature ganglioneuroma with sparse spindle cells and, occasionally, ganglionoid cells. Immunohistochemical results showed that tumor cells expressed Vimentin (3+), S100 (2+), and SOX10 (2+) and were negative for SMA, EMA, CD-117, and the Ki-67 proliferation index was less than 1% (Figure 3). Finally, the patient underwent an open resection. The mass was carefully separated from the inferior vena cava and multiple small feeding vessels were ligated. Gross pathologic examination showed a large nodular mass measuring 21 cm × 14 cm × 11 cm in size (Figure 4). Histopathological analysis confirmed the diagnosis. The patient was followed for 24 months and no tumor recurrence occurred.

3 | DISCUSSION AND CONCLUSIONS

GN is a rare and slow-growing tumor that originates in the sympathetic neural crest and consists of mature ganglion cells, Schwann cells, nerve fibers, and a rich

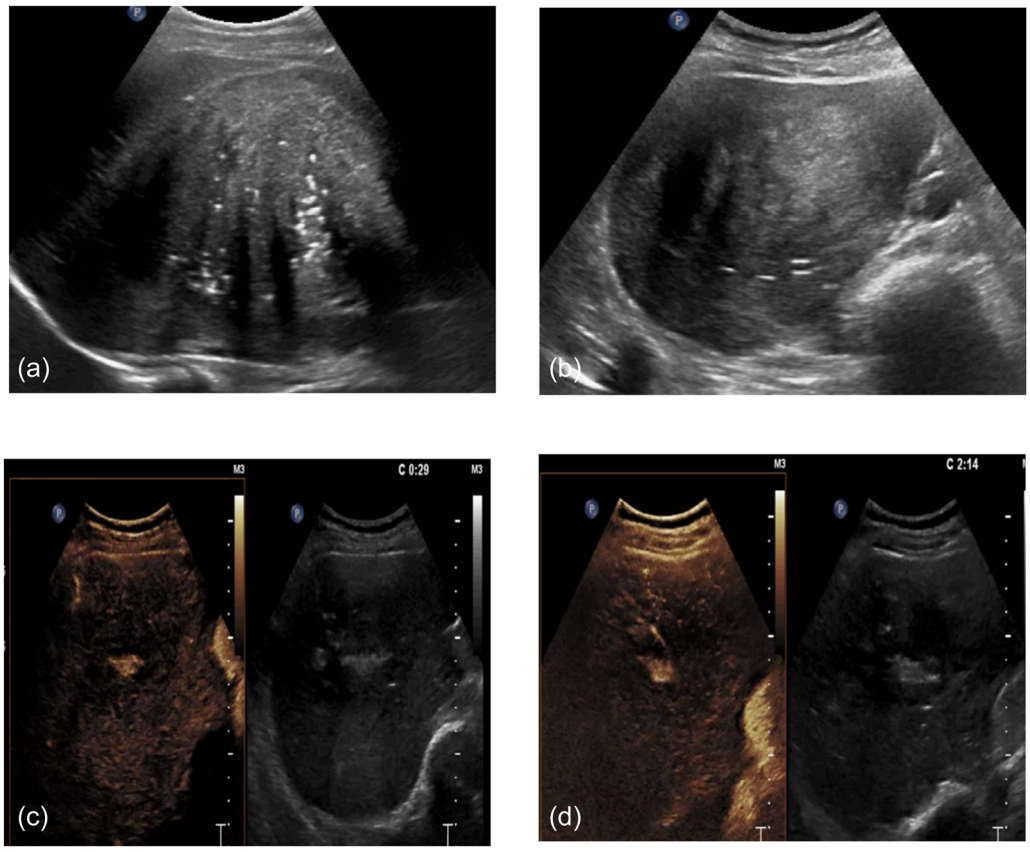

FIGURE 1 Multimodal ultrasound discovery of the giant mass. (a) B-mode ultrasound longitudinal scan shows a homogeneous and hypoechoic mass with multiple punctate calcifications; (b) the short axis of the transversal scan shows that the mass has well-defined edges; (c) CEUS shows mild and heterogeneous enhancement of the arterial phase; and (d) CEUS continues to enhance during the delayed phase. CEUS, contrast-enhanced ultrasonography.

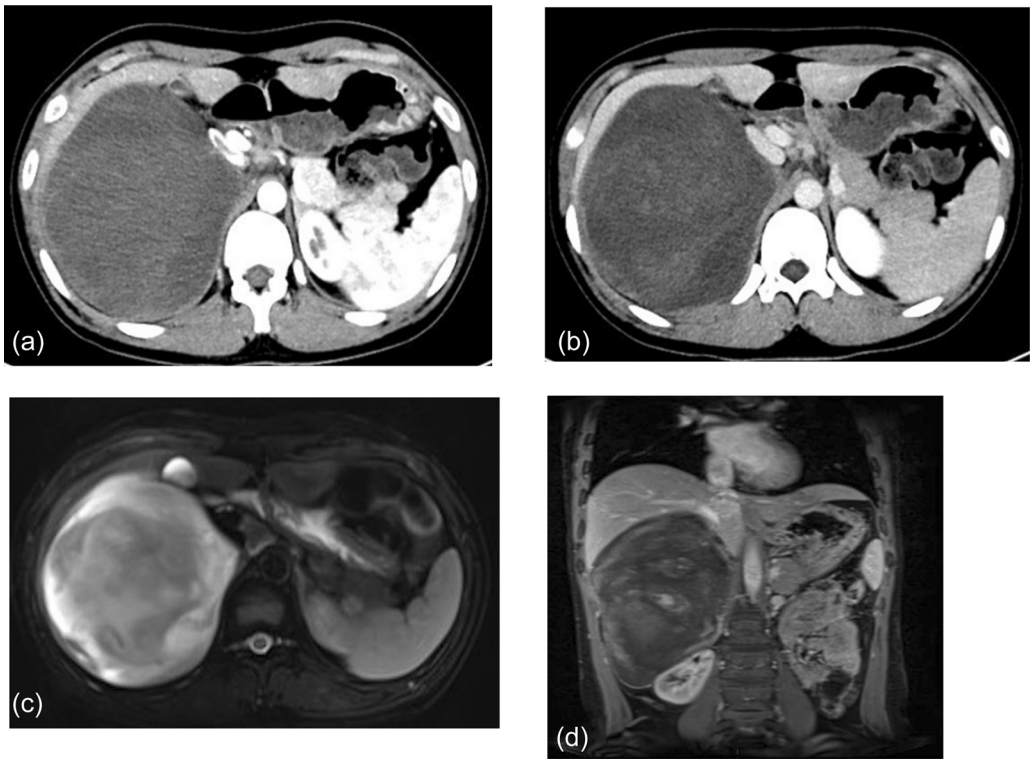

FIGURE 2 CT and MRI images of the patient. (a) Contrast-enhanced CT shows that the hypodense mass has no enhancement in the arterial phase; (b) CT scan reveals the mass showing inhomogeneous flocculent enhancement in the venous phase; (c) MRI shows a heterogeneous high T2 signal; and (d) Contrast-enhanced MRI shows mild enhancement in the delayed phase and anterior displacement of the inferior vena cava and portal vein. CT, computed tomography; MRI, magnetic resonance imaging.

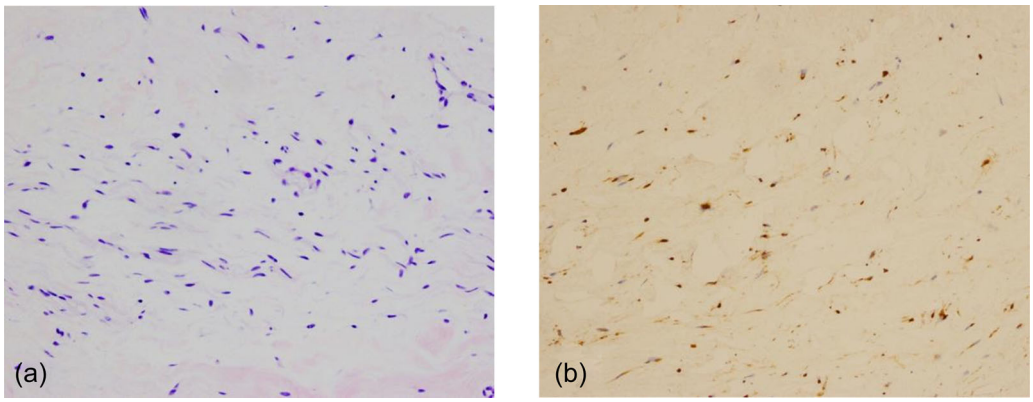

FIGURE 3 Pathological features detected by ultrasound-guided core needle biopsy. (a) Histopathology shows the sparseness of spindle-shaped cells in the mucus matrix (H&E, ×200); and (b) immunohistochemical staining shows tumor interstitial cells expressing Schwann cell marker S-100 (++, Envision, ×200). H&E, hematoxylin and eosin.

mucus matrix. Most GNs may grow in a creeping pattern along the surrounding organ space, with vascular embedding but no invasion [3], reflecting their benign biological behavior. Immunohistochemical analysis of S-100, NSE, and other neurogenic markers may be positive. Retroperitoneal GN is more common in individuals younger than 20 years of age at initial diagnosis [4]. Tumors are usually discovered incidentally on physical examination and have nonspecific clinical symptoms. As small GN grows, hormones (e.g., catecholamines, vasoactive intestinal peptides, etc.) may be released, leading to hypertension, diarrhea, and hyperhidrosis [2]. Serum neuron-specific enolase (NSE), ferritin, lactate dehydrogenase, 24-h urine vanillylmandelic acid (VMA), and other neurogenic tumor markers may remain normal [5]. Therefore, the clinical diagnosis

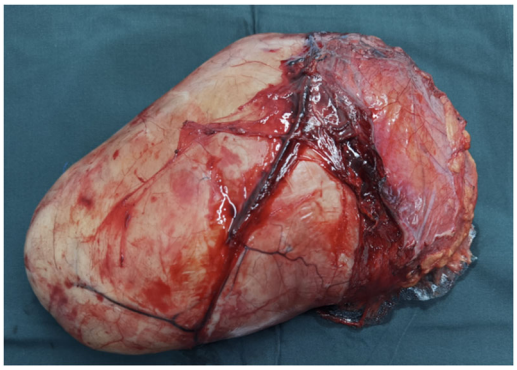

FIGURE 4 Gross pathological examination shows a large nodular mass of 21 cm × 14 cm × 11 cm.

of retroperitoneal GN remains a challenge due to the low incidence and lack of specific clinical manifestations and laboratory indicators.

US plays an important role in diagnosing retroperitoneal GN. In conventional US, GN presents as a homogeneous, well-defined, and hypoechoic retroperitoneal mass with mild vascularity. Punctate calcifications are present in one-third of cases [6], which is consistent with our case. On CEUS, GN can manifest as progressive mild enhancement, which is associated with an abundant mucus matrix, relatively few ganglion cells, and interstitial vascular tissue. CT is beneficial in showing the relationship between the tumor and adjacent structures. It is usually a near-cystic density, oval mass with scattered punctate calcifications but no necrosis. MRI T2WI is often characterized by hyperintensity and a "whorled sign" due to the large amount of mucus matrix mixed with Schwann cells and collagen fibers [6]. Both contrast-enhanced CT and MRI often show a delayed progressive enhancement pattern [6]. Ultrasound-guided biopsy further confirms the diagnosis of the tumor. The Chinese Expert Consensus on the Treatment of Retroperitoneal Tumors (Edition 2019) [7] strongly recommends image-guided core needle biopsy rather than fine-needle aspiration for tumors that are difficult to resect or differentiated from other diseases. Therefore, when a retroperitoneal solid mass is detected in the conventional US, multimodal ultrasound indicates benign neurogenic tumors, and ultrasound-guided core needle biopsy combined with immunohistochemical markers can be performed to obtain a correct preoperative diagnosis. Complete surgical resection is the best treatment for GN [8], and laparoscopic surgery may be considered in patients with relatively simple anatomical structures and tumors less than 10 cm in diameter. Chemotherapy or radiotherapy is of little value [9]. Due to the risk of late recurrence of GN, long-term follow-up is necessary. In our patient, the tumor was larger than 20 cm. Given the safety of the operation and complete resection, laparotomy was performed, with good results.

We present this case to highlight the value of multimodal ultrasound imaging in the diagnosis of retroperitoneal solid tumors. The difficulties, in this case, are the low incidence of retroperitoneal GN, complex histological types, lack of specificity of clinical manifestations, and confusion in imaging features. First, in terms of localization, it is helpful to determine whether the tumor is located in the abdominal cavity or behind the peritoneum by real-time and multisegment scanning of US. When the patient is asked to breathe and change body position, we can assess whether there are "peak signs" and "hanging tumor signs," or to observe whether the tumor compresses the retroperitoneal organs or blood vessels, and whether the liver is separated from the right kidney and the spleen from the left kidney. Second, conventional US can be used to observe tumor size, depth, shape, boundaries, internal echo, calcification, growth pattern, relationship with the surrounding organs and blood vessels, and blood flow signals. Moreover, CEUS can also be included to observe microvascular perfusion inside the tumor to obtain more diagnostic clues. Meanwhile, combined with clinical features such as age and symptoms, CT and MRI imaging results can further enhance the confidence of ultrasound diagnosis. Finally, ultrasound-guided core needle biopsy combined with immunohistochemistry is useful in confirming the diagnosis and individualizing treatment strategies.

AUTHOR CONTRIBUTIONS

Li Feng: Data curation (lead); formal analysis (lead); software (lead); writing—original draft (lead); writing—review and editing (equal). **Yong Wang**: Conceptualization (lead); funding acquisition (lead); investigation (lead); methodology (lead); project administration (lead); resources (lead); supervision (lead); validation (lead); visualization (lead); writing—review and editing (equal).

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CONFLICT OF INTEREST STATEMENT

Professor Yong Wang is a member of the *Cancer Innovation* Editorial Board. To minimize bias, he was excluded from all editorial decision-making related to the acceptance of this article for publication. The remaining author declares no conflict of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICS STATEMENT

The study protocol was approved by the Ethics Committee of the Cancer Hospital of the Chinese Academy of Medical Sciences (NCC2016YZ-15).

INFORMED CONSENT

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

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