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Case Report

Retroperitoneal ganglioneuroma simulating lymphoma: An unusual case presentation ☆

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ABSTRACT

Ganglioneuromas of the retroperitoneum are rare cases of benign retroperitoneal tumors that arise from autonomic nerve fibers. Typically remaining asymptomatic even after growing very large, retroperitoneal ganglioneuromas are often discovered incidentally. Given the rather nonspecific, well-defined, smooth, or lobulated characteristics similar to most neurogenic tumors, retroperitoneal ganglioneuromas are sometimes difficult to diagnose. Surgical resection is the preferred treatment option intended to limit the chance of recurrence or malignant transformation; however, surgery can be restricted by local extension, such as encasement of blood vessels. In this article, we report the case of a 49-year-old female diagnosed with a retroperitoneal ganglioneuroma presenting as abdominal pain and fullness. We focus on the use of radiological imaging modalities to coordinate with surgical pathology for an optimized diagnosis and treatment plan.

Introduction

Ganglioneuromas (GNs) are rare benign tumors that arise from mature Schwann cells, ganglion cells, and nerve fibers, capable of presenting anywhere along the paravertebral sympathetic plexus [1,2]. Primarily occurring in the retroperitoneal space, GNs constitute only a small percentage of retroperitoneal tumors, between 0.72% and 1.6%, and have a reported incidence of one per million population [2,3]. Retroperitoneal GNs are usually nonfunctional and asymptomatic until large enough to cause symptoms by local expansion and compression of nearby structures [4,5]. Patients most often present with lower abdominal/back pain or with concern of a distended abdomen with a palpable mass. The rarity and nonspecific clinical nature of GNs make this tumor difficult to discern from other neurogenic benign or malignant masses, especially lymphoma when it appears to infiltrate around or encase vessels. This requires the usage of radiological imaging modalities with the coordination of pathological findings to suggest specific diagnoses for management and preoperational planning. In this article, we report the case of a 49-year-old female diagnosed with a retroperitoneal GN that remained inoperable due to its encasement of major blood vessels.

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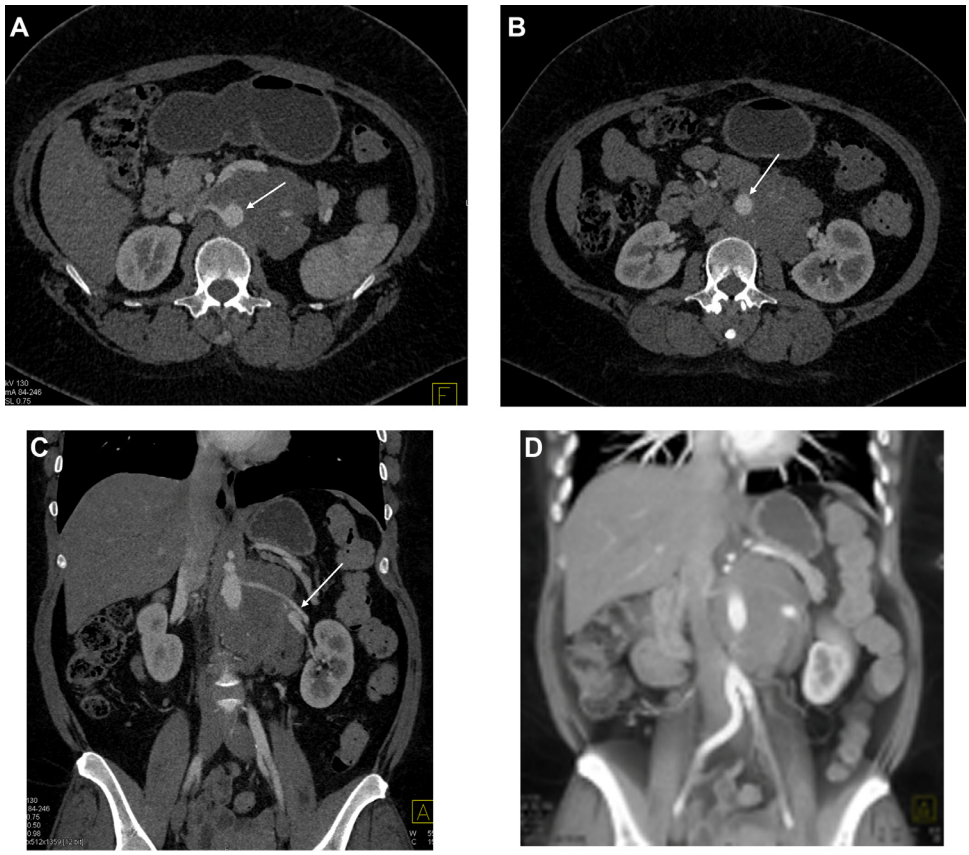

Fig. 1 – 49-year-old female with symptoms of nausea, vomiting, abdominal pain, and fullness. (A-B) Axial contrast-enhanced CT scans demonstrate a 11.7 x 8.3 x 6.4 cm low attenuation mass (30 HU) encasing the aorta (arrow) and renal arteries. No evidence of bone involvement is seen. (C) Coronal CT scans demonstrate the displacement of the left kidney and the vascular encasement including the left renal artery (arrow). (D) 3D mapping with volume rendering defines the extent of the tumor and the lower density nature of the mass.

Case presentation

A 49-year-old female with no significant past medical history presented to an outside institution with symptoms of nausea and vomiting, complaining of abdominal pain and fullness. Physical evaluation of the abdomen was otherwise inconclusive, revealing no tenderness or distension. To further investigate, the patient was scheduled for radiological imaging of the chest and abdomen.

A chest computed tomography (CT) identified no significant thoracic abnormality or adenopathy within the chest cavity. A subsequent CT of the abdomen and pelvis with intravenous (IV) contrast (Fig. 1; A-D) revealed a 11.7 cm x 8.3 cm x 6.4 cm mass, centered in the left retroperitoneum. The mass encased the upper abdominal aorta, lumbar arteries, origin of the right renal artery, left renal artery, and anteriorly displacing and narrowing the left renal vein. No association with either adrenal gland was noted. Initial impressions were suggestive of lymphoma, neurogenic tumor, or sarcoma. On positron emission tomography CT (PET–CT), the retroperitoneal mass showed heterogeneous moderately increased metabolic activity with SUV max of 5.2. A diagnostic laparoscopy and surgical biopsy showed neural tissue with ganglion cells, consistent with ganglioneuroma. Complete surgical resection of the GN was precluded by the encasement of major blood vessels. Patient subsequently underwent serial surveillance imaging in 6–12-month intervals.

This patient later presented to the ER with concern of right upper quadrant (RUQ) pain that radiated from her epigastrium and worsened until presentation. During the visit, CT and MRI scans were again taken to evaluate the abdominal cavity. The CT revealed no substantial changes in the extent of the retroperitoneal soft tissue mass. The MRI (Fig. 2; A-D) showed a 10.5 cm lobulated mass with mild heterogeneity and progressive enhancement, consistent with the previously diagnosed ganglioneuroma. Patient had acute cholecystitis from gallstones and underwent subsequent cholecystectomy.

Discussion

Here, we present the rare case of a benign retroperitoneal GN, arising from the ganglion cells of the paravertebral sympathetic plexus [3]. Ganglioneuromas are mature and well-differentiated subtypes of neuroblastic tumors and can be multiple and/or associated with other types

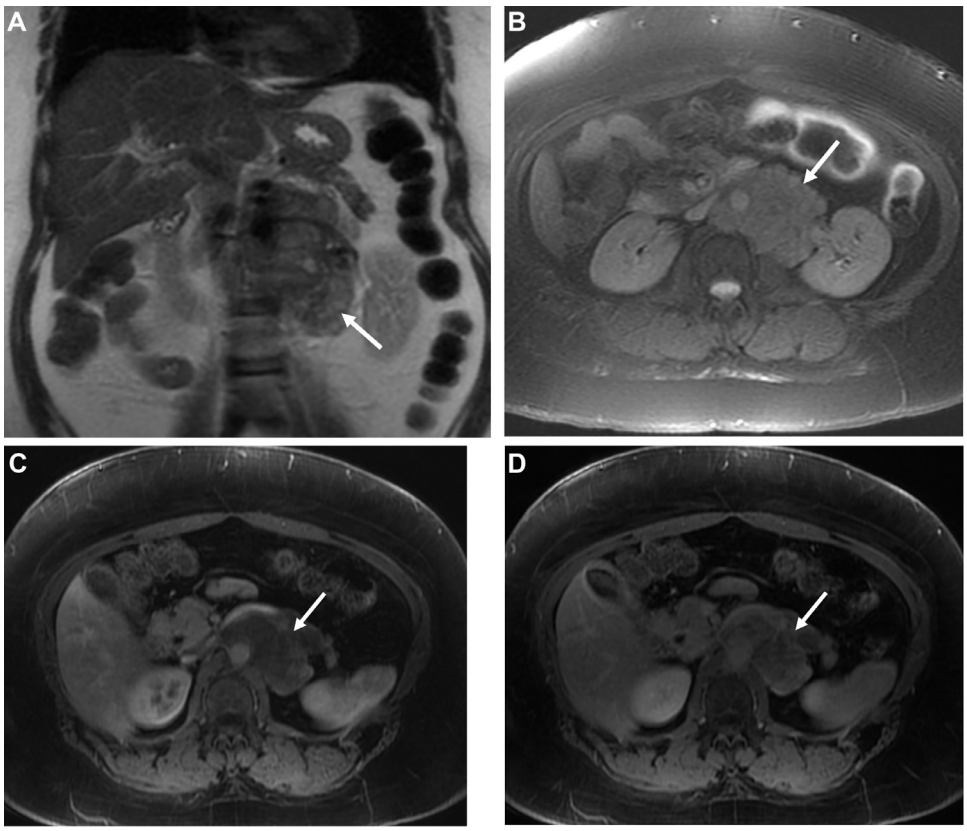

Fig. 2 – (A) Coronal T2-weighted MR and (B) axial fat-saturated T2-weighted MR images also demonstrate a lobulated left retroperitoneal mass with intermediate heterogeneous signal intensity (arrows) and vascular encasement. (C-D) Axial postcontrast T1-weighted MR in the arterial phase (C) and portal venous phase (D) show heterogeneous progressive enhancement of the left retroperitoneal mass (arrows).

of neural/neuroendocrine neoplasms of the periphery [5,6]. Retroperitoneal GNs are sometimes difficult to differentiate from malignant neuroblastic tumors preoperatively and rely on pathologic coordination for conclusive diagnosis [1,4,7]. Rare occurrences of spontaneous malignant transformations have arisen from retroperitoneal GNs in the form of nerve sheath tumors, however, generally GNs are slow growing and noncancerous [8,9]. When these GN tumors are infiltrating, as in our case, concern for lymphoma is also in the differential diagnosis.

A retrospective study by Xiao et al. [6] describes the demographics and presentations of 32 patients diagnosed with retroperitoneal GN. This tumor was discovered in individuals between the age of 15-62 years with a mean of 35 years and a sex ratio of 1:3 male to female, however, the ratio is closer to 1:1 in other published studies [9]. 65.6 % of patients were asymptomatic, and lesions were found incidentally. The remaining 34.4 % presented with a variety of symptoms including abdominal pain, weight loss, peripheral numbness, and pain from nerve compression [6]. Infrequently, GNs produce symptoms like diarrhea, hyperhidrosis, flushing, and hypertension from the production of catecholamines and vasoactive intestinal peptides, despite being endocrine-inactive [9]. Often GNs surround major blood vessels, however, no indications of complete obstruction or invasion of luminal space have been reported [2]. As in our case, abdominal pain and fullness likely can be attributed to the displacement of surrounding structures and organs.

Radiological imaging is essential for the precise localization, diagnosis, and preoperative planning of GNs, used to evaluate the extent of the tumor including organ of origin, vascular encasement, adenopathy, and calcification [10]. Subsequent coordination with surgical pathology optimizes the diagnosis and directs the following treatment plan. Nonenhanced CT imaging of GNs usually reveals a well-circumscribed oval, crescentic, or lobulated mass of homogeneous or mildly heterogeneous low attenuation, and sometimes discrete and punctate calcifications in 20 % of retroperitoneal GN cases. Since there are various degrees of overlap between the imaging appearance of GNs and malignant neurogenic tumor types including ganglioneuroblastomas and neuroblastomas, the presence of calcification is considered for differential diagnosis with neuroblastoma calcifications being described as amorphous and coarse [2,7,11]. While there was no calcification in our case, CT images of our patient revealed a large lobulated well-circumscribed hypodense mass that were typical of ganglioneuromas.

GNs typically demonstrate soft tissue density with hypo- to iso-intensity in T 1W1, iso-to hyper-intensity in T2W1, and hyperintensity in DWI on MRI, mostly reporting enhancement

[6]. A 5-case study completed by Saadi et al. [12] describes the retroperitoneal GN as homogeneously hypointense on T1-weighted sequences and iso- or hyperintense on T2-weighted sequences. The GN T2 signal intensities depend on the proportion of myxoid stroma to the cellular component and the amount of collagen fibers in the tumor [12]. Tumors with high ratios of cellular components and collagen fibers to myxoid stroma displayed intermediate – hyper T2 signal intensities, while the reverse ratio displayed a distinctly high signal intensity on the T2-weighted images [2]. Enhancement following gadolinium injection was nonspecific, varying between absent, discrete, heterogeneous, and intense. Studies have explored the utility of FDG-PET scanning in differentiating benign and malignant retroperitoneal masses, and while our case showed an SUVmax above the threshold of 3.13 ± 1.53 determined by Liu et al. in their review and helped the diagnosis, FDG-PET has not been shown to effectively discriminate between benign and malignant lesions [13].

The benign nature of the retroperitoneal ganglioneuroma tends to favor a positive prognosis, however, complete surgical resection is always considered to prevent recurrence and the small chance of malignant transformation [12]. Frequently, surgical resection is impossible due to vascular encasement. In the case of our 49-year-old patient, encasement of the upper abdominal aorta and renal arteries restricted the ability for a complete surgical resection necessitating serial imaging surveillance.

Conclusion

Retroperitoneal ganglioneuromas are rare presentations of benign retroperitoneal tumors arising from the paravertebral sympathetic plexus, comprising roughly 0.72%-1.6% of all retroperitoneal masses. Radiological imaging is important for visualizing the extent of the mass, however, differentiating from other neuroblastic tumors may be difficult, requiring coordination with surgical pathology for an optimized diagnosis and treatment plan. Retroperitoneal GNs have an overall optimistic prognosis but tend to encase surrounding major blood vessels rendering them inoperable and requiring extended follow-up imaging.

Patient consent

The patient reported in the manuscript signed the informed consent/authorization for participation in research, which includes the permission to use data collected in future research projects such as the presented case details and images used in this manuscript.

REFERENCES

[1] Moriwaki Y, Miyake M, Yamamoto T, Tsuchida T, Takahashi S, Hada T, et al. Retroperitoneal ganglioneuroma: a case report and review of the Japanese literature. Intern Med 1992;31(1):82–5. doi:10.2169/internalmedicine.31.82.

[2] Rha SE, Byun JY, Jung SE, Chun HJ, Lee HG, Lee JM. Neurogenic tumors in the abdomen: tumor types and imaging characteristics. Radiographics 2003;23(1):29–43. doi:10.1148/rg.231025050.

[3] Geoerger B, Hero B, Harms D, Grebe J, Scheidhauer K, Berthold F. Metabolic activity and clinical features of primary ganglioneuromas. Cancer 2001;91(10):1905–13. doi:10.1002/1097-0142(20010515)91:10⟨1905::aid-cncr1213⟩3.0.co;2-4.

[4] Bove KE, McAdams AJ. Composite ganglioneuroblastoma. An assessment of the significance of histological maturation in neuroblastoma diagnosed beyond infancy. Arch Pathol Lab Med 1981;105(6):325–30.

[5] Zare S, Parvin M, Ghohestani SM. Retroperitoneal ganglioneuroma. Urol J 2008;5(4):232.

[6] Xiao J, Zhao Z, Li B, Zhang T. Primary retroperitoneal ganglioneuroma: a retrospective cohort study of 32 patients. Front Surg 2021;8:642451. doi:10.3389/fsurg.2021.642451.

[7] Ichikawa T, Ohtomo K, Araki T, Fujimoto H, Nemoto K, Nanbu A, et al. Ganglioneuroma: computed tomography and magnetic resonance features. Br J Radiol 1996;69(818):114–21. doi:10.1259/0007-1285-69-818-114.

[8] Ghali VS, Gold JE, Vincent RA, Cosgrove JM. Malignant peripheral nerve sheath tumor arising spontaneously from retroperitoneal ganglioneuroma: a case report, review of the literature, and immunohistochemical study. Hum Pathol 1992;23(1):72–5. doi:10.1016/0046-8177(92)90015-u.

[9] Kitazawa M, Matsuhashi N, Imai T, Iwata Y, Takahashi T, Yoshida K. Total laparoscopic excision of retroperitoneal ganglioneuroma: a case report. Int J Surg Case Rep 2021;83:106053. doi:10.1016/j.ijscr.2021.106053.

[10] Singh J, Kr Priyadarshi V, Kumar Pandey P, Kr Vijay M, Kumar Pal D, Kundu A. Retroperitoneal ganglioneuroma. APSP J Case Rep 2013;4(1):8.

[11] Otal P, Mezghani S, Hassissene S, Maleux G, Colombier D, Rousseau H, et al. Imaging of retroperitoneal ganglioneuroma. Eur Radiol 2001;11(6):940–5. doi:10.1007/s003300000698.

[12] Saadi A, Chebbi S, Mokadem S, Kacem LBH, Chakroun M, Slama MRB. Retroperitoneal ganglioneuroma: a five-case series from a single Tunisian center. Int J Surg Case Rep 2023;111:108840. doi:10.1016/j.ijscr.2023.108840.

[13] Ning Liu D, Wu Li Z, Yue Wang H, Zhao M, Zhao W, yi Hao C. Use of 18F-FDG-PET/CT for retroperitoneal/intra-abdominal soft tissue sarcomas. Contrast Media Mol Imaging 2018;2018:2601281. doi:10.1155/2018/2601281.