SAGE Open Medical Case Reports
Volume 12: 1–5

DOI: 10.1177/2050313X241252744
journals.sagepub.com/home/sco
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Large retroperitoneal ganglioneuroma revealed by a left ovarian endometrioma: A case report

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Abstract

Uncommon in nature, retroperitoneal ganglioneuroma represents a neuroblastic benign tumor, predominantly manifesting in young adults, with a notable predilection for females. Often asymptomatic, the condition is frequently diagnosed incidentally due to delayed growth. Clinical manifestations arise primarily from the compression exerted on neighboring organs and vessels. The exclusive curative recourse lies in surgical intervention, underscoring the challenging task of achieving complete tumor excision, particularly when the ganglioneuroma attains considerable development and encapsulates significant retroperitoneal vessels. In this instance, we elucidate a case involving a 33-year-old woman, who had previously undergone a triple valve replacement due to rheumatic valvular disease, presenting persistent pelvic pain, unearthing a substantial asymptomatic retroperitoneal ganglioneuroma concomitant with an ovarian endometrioma. A laparotomy procedure was conducted, and to achieve a comprehensive excision of the mass, a meticulous intratumoral circular dissection of the prominent vessels, notably the superior mesenteric artery and celiac trunk, was undertaken. No local recurrence has been reported, 6 months after surgery. The significance of an experienced and well-trained surgical staff is underscored in addressing the complexities associated with this condition.

Keywords

Retroperitoneal ganglioneuroma, ovarian endometriosis, complete tumor resection

Date received: 30 November 2023; accepted: 18 April 2024

Introduction

Ganglioneuromas (GNs) are uncommon benign tumors originating from the neural crest, classified as extracranial neuroblastic tumors. Predominantly found in the retroperitoneum (32%–52%) adjacent to the paravertebral sympathetic chain, they infrequently arise from the adrenal medulla. These tumors exhibit the presence of developed Schwann cells, ganglion cells, and nerve fibers.[1,2] GNs are characterized by a female predilection and frequently observed in children and young adults.[3] They are typically asymptomatic, whereas the majority of cases lack definitive clinical manifestations, with incidental discovery occurring through radiological imaging while investigating unrelated pathologies.[4] Symptomatic presentation ensues as a consequence of the tumor's progressive growth and compression of neighboring organs and vasculature.[2] A conclusive diagnosis relies on histopathological examination, which typically reveals a blend of ganglion cells and Schwann cells without immature elements.[2,4] Surgical intervention remains the sole curative option, with the efficacy contingent upon the size and regional expansion of the tumor.[2,3]

We report a case of a young woman with an enormous retroperitoneal GN that intricately encircled the abdominal arteries at their origin, rendering the surgical procedure

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intricate and fraught with considerable risk. The incidental discovery of this massive asymptomatic tumor occurred during imaging conducted for chronic pelvic pain, initially due to an endometriotic ovarian mass misdiagnosed as an ovarian dermoid cyst. Confirmation of both left ovarian endometrioma and GN diagnoses was achieved through histological analysis of the specimens.

Case presentation

A 33-year-old woman, who had undergone a triple valve replacement for rheumatic valvular disease and was receiving acenocoumarol, presented with a 12-month history of paroxysmal and intermittent left iliac fossa pain, exacerbating during menstruation and accompanied by an estimated weight loss of 10 kg. Physical examination revealed normal findings, except for heightened sensitivity in the left iliac fossa and hypogastrium. The patient exhibited anemia with a hemoglobin level of 9.9 g/dL, a prothrombin time of 40%, and an international normalized ratio (INR) of 1.5. The rest of the blood profile, including tumor markers (carcinoembryonic antigen at 2.21 ng/ml, carbohydrate antigen 19-9 (CA19-9) at 24.3 U/mL, and cancer antigen 125 (CA 125) at 34.4 U/mL), was within normal limits.

Initially, an abdominopelvic ultrasound was conducted, complemented by an angiographic computed tomography (CT) scan, revealing a well-encapsulated retroperitoneal mass on the right side, clear and lobed outline projecting onto the epigastrium and right hypochondria. The mass measured $118 \times 92 \times 120$ mm and exhibited very low heterogeneous density. It made contact with the pancreas, spleno-mesenteric trunk, and portal trunk anteriorly, while sliding posteriorly between the abdominal aorta and the inferior vena cava (IVC). The mass extended into the right anterior perirenal space, encompassing the homolateral renal artery and contacting the homologous renal vein. It overflowed the midline, enclosing the celiac trunk (CeT) and the superior mesenteric artery (SMA), while making contact with the left adrenal gland, splenic vein, and left renal vein (LRV). Externally, the mass intimately interacted with the liver and gallbladder, pushing the right adrenal gland against the VII segment of the liver. It encased the IVC over 180° of its circumference without compromising its opacification. No intra- or retroperitoneal lymphadenopathy was observed (Figure 1). In the pelvic space, a left ovarian cystic tumor was identified, hypodense with a thin wall, containing discreetly hyperdense sloping material, indicative of an ovarian dermoid cyst classified as O-rads 2 (on ultrasound; Figure 2).

To obtain a definitive diagnosis, an upper gastrointestinal echo-endoscopy with biopsy of the retroperitoneal mass was performed. Pathological examination revealed a tumor proliferation characterized by a schwannoma stroma containing fusiform cells with elongated nuclei. Scattered mature ganglionic cells were also present, resembling the histological appearance of ganglioneuroma.

Despite the apparent difficulty or even impossibility of complete resection based on CT data, surgical abstention was deemed risky due to the potential for wider extension, compression complications, and subsequent riskier resection afterward. Moreover, the presence of the pelvic mass prompted surgical intervention. In light of the patient's significant medical history, requisite preoperative measures were undertaken. These included discontinuation of acenocoumarol, necessitating a transition to enoxaparin sodium at a dosage of 6000 UI administered twice daily, with the cessation of administration 12 h prior to the scheduled procedure to mitigate the risk of bleeding. An INR test was conducted to assess the coagulation status. A comprehensive cardiac assessment was carried out, encompassing an electrocardiogram and a transthoracic ultrasound. Additionally, a thorough hematological evaluation was conducted on the day preceding the surgical intervention. Subsequently, a requisition for packed red blood cells and platelets was made on the day of the surgery to ensure preparedness for potential transfusion requirements. In anticipation of complications, provision was made for the patient to be accommodated in the surgical intensive care unit.

Abdominal exploration through xyphopubic laparotomy confirmed the CT scan findings, revealing the giant retroperitoneal mass in contact with adjacent organs and enveloping large retroperitoneal vessels. A complete resection necessitated the opening of the tumor in contact with large vessels and a circular dissection of the CeT and the SMA, posing a risk of damaging the surrounding nerve plexus.

The exploration further identified a left ovarian cyst, from which brownish fluid was extracted, more suggestive of endometriosis than dermoid cysts, leading to cystectomy. The postoperative course was uneventful, and the patient was discharged on the sixth day with subsequent removal of drains. The final diagnosis was confirmed through histopathological examination of the specimens, revealing a synchronous retroperitoneal ganglioneuroma and a left ovarian endometrioma (Figure 3). The patient was subsequently referred to a gynecologist for endometriosis management. As of 6 months post-surgery, no local recurrence has been reported.

Discussion

Retroperitoneal GNs are distinctly differentiated benign tumors originating from the retroperitoneal sympathetic ganglia, exhibiting an exceptional rarity with an incidence of one per million in the general population.[2] Classified as extracranial neuroblastic tumors along with neuroblastoma and ganglioneuroblastoma, GNs have the capacity to develop in various anatomical locations, with the posterior mediastinum and retroperitoneum being the predominant sites at 32%–52% and 39%–42%, respectively.[2] Furthermore, these tumors can manifest in the neck and adrenal glands, with a notable inclination toward a higher incidence among

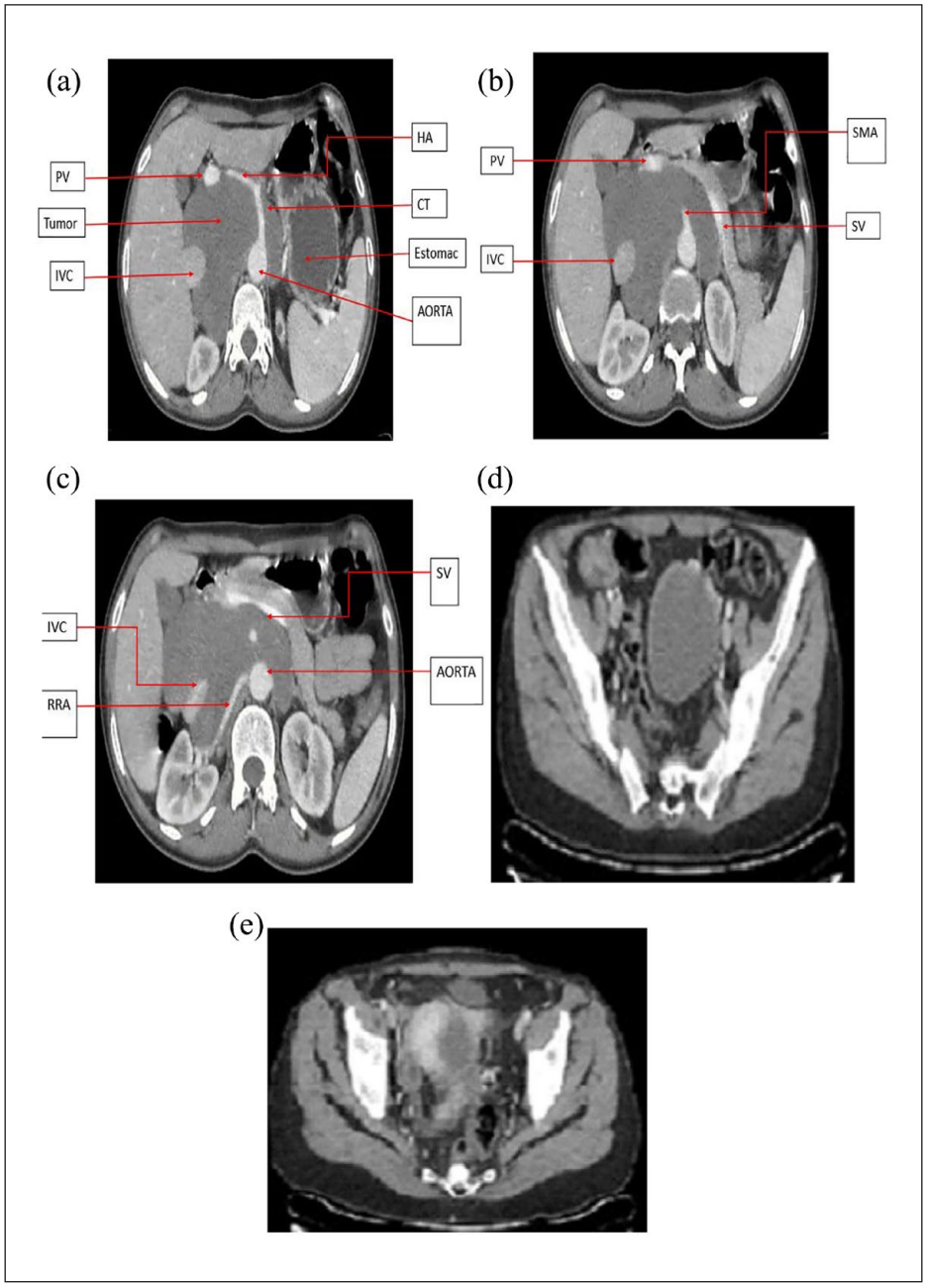

Figure 1. Computed tomography (CT) angiography of abdominopelvic imaging in retroperitoneal ganglioneuroma. (a) Giant retroperitoneal ganglioneuroma right sided under liver, slides between and encompassing the inferior vena cava (IVC) and aorta, it pushes forward the portal trunk and encases the celiac trunk, the aorta and the IVC more than 180°. (b) Retroperitoneal ganglioneuroma (RGN) encases the right renal artery at its origin and comes in contact with the right renal vein. (c) RGN gets in contact with the forward pancreas and splenic vein. Back, it is extended to anterior pararenal and perirenal space. It encases the superior mesenteric artery at its origin. (d) and (e) CT angiography of abdominopelvic imaging; an ovarian cystic mass pushes right to the uterus and measures 7 cm. HA: hepatic artery; PV: portal vein.

women.[3] Multiple endocrine neoplasia II or neurofibroma II are recognized as potential risk factors for the development of this benign tumor.[3]

Typically asymptomatic and characterized by slow growth, GNs are frequently incidentally discovered during radiological imaging conducted for unrelated disorders. Nevertheless, site-specific symptoms may manifest as the tumor enlarges, compressing and displacing adjacent organs and vessels, with prevalent symptoms including abdominal back discomfort and abdominal distension.[5,6] While GNs are

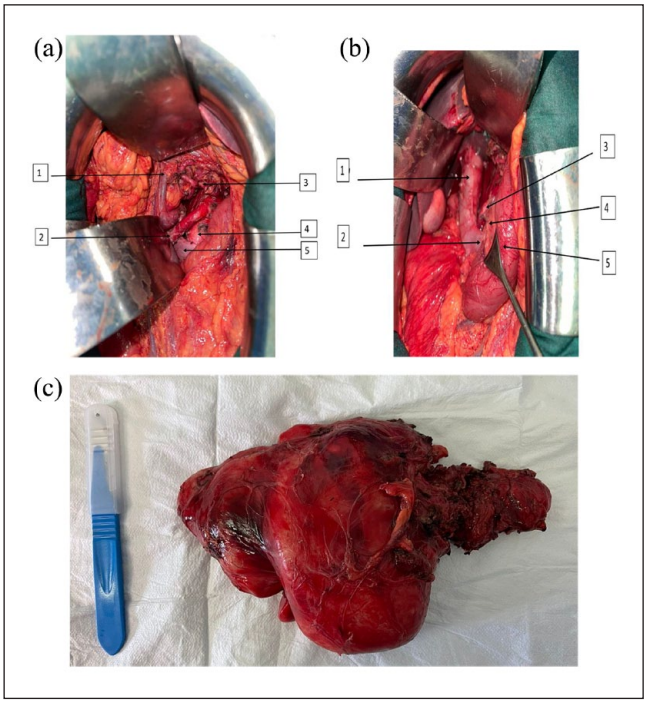

Figure 2. (a) Perioperative view after total resection of Ganglioneuromas (GN) showed the release of all retroperitoneal vascular structure 1: inferior vena cava (IVC) 2: superior mesenteric artery (SMA) 3: celiac trunk (CeT) 4: Aorta 5: left renal vena (LRV). (b) Perioperative view after complete resection of RGN demonstrate the release of all retroperitoneal vascular structures. 1: IVC. 2: LRV. 3: SMA. 4: aorta. 5: stomach. (c) Image of the final surgical specimen of the GN.

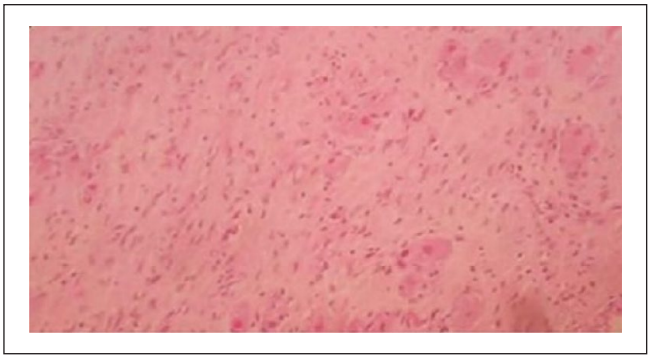

Figure 3. Ganglioneuroma with admixture of ganglion cells and mature Schwann cells.

generally nonsecretory tumors, some instances involve the secretion of catecholamine and/or vasoactive intestinal polypeptides, leading to symptoms such as hypertension and diarrhea.[6,7]

Radiological imaging, particularly on CT scans, often depicts GN as a homogeneous, well-defined oval mass with little or minimal arterial phase enhancement and gradual mild enhancement in the delayed phase. Punctate calcifications are observed in 20% of cases. Magnetic resonance imaging reveals a mass that is isointense or hypointense to the spinal cord on T1-weighted images, hyperintense on T2-weighted images, and heterogeneous on contrast-enhanced T1-weighted images. Imaging methods are crucial for surgical planning, providing insights into the tumor's size, origin organ, local expansion, and conjunction with surrounding organs and arteries.[2,4,6] However, it is imperative to note that the misinterpretation of a benign neuroblastic tumor as a sarcoma by a CT scan has been documented, underscoring the inadequacy of imaging features in providing an accurate diagnosis for this condition.

The diagnosis may be aided by preoperative cytology employing fine needle aspiration by upper gastrointestinal echo-endoscopy, as in our case. Besides, the presence of adipose cells in these tumors, previously reported in the literature and due to the lack of distinguishing characteristics, may make it difficult to differentiate between GNs and other aggressive tumors using cytology that leads to delay a proper treatment.[6] Consequently, histopathological examination of the specimen remains the sole specific test for a conclusive diagnosis of GN, identifying an admixture of ganglion cells and Schwann cells without immature elements.[6,7]

The only recommended therapeutic approach is complete tumor resection, given its efficacy and low recurrence rate. However, the procedure becomes riskier when the GN extensively expands, compressing surrounding organs with vascular encasement. The main complications associated with such operations include bleeding, ischemia, and notably neurological dysfunction, such as temporary or treatment refractory diarrhea, which may occur after extensive dissection of the SMA.[8] In the presented case, postoperative diarrhea was not observed. Preoperative radiological imaging plays a crucial role in determining the adequacy of the therapeutic approach by assessing the tumor's size, connection to adjacent organs, and the likelihood of total resection.[6,9]

Incomplete resection remains a viable option, especially when the residual tumor measures less than 2 cm. Existing studies suggest that this approach does not pose an increased risk of progression. For that reason, a rationale for reducing the tumor may be considered if the tumor is symptomatic or hormonally active with vaste extension and a higher risk of morbidity.[10] This consideration is particularly relevant given the benign and slow-growing nature of these tumors. In situations where surgery is declined by the patient or is associated with potential risks, an abstinent approach with surveillance may be a viable option, especially since GNs are often asymptomatic and stable.[7,10] In the case presented, a thorough deliberation led to the selection of a therapeutic surgical approach. The procedure posed technical challenges due to the considerable enlargement of the GN, encasing major retroperitoneal arteries, and proved demanding for the surgical team.

Nevertheless, the positive outcomes observed in our patient endorse the suggestion for fellow surgeons to contemplate complete surgical excision whenever feasible for such large benign tumors.

Conclusion

GNs, characterized by their slow growth and lack of hormonal functionality, are typically incidentally identified. Notably, these benign tumors exhibit a distinctive trait of enveloping retroperitoneal organs, particularly significant retroperitoneal arteries, without infiltrating them. This peculiarity underscores the challenges associated with achieving a comprehensive excision of the tumor. Moreover, considering the extended period of recurrence-free disease subsequent to incomplete tumor excision, the latter option may serve as an acceptable alternative. Ultimately, the concurrent occurrence with pelvic endometrioma is unprecedented and appears to be a fortuitous circumstance, but the clinical manifestation in our case serves as a crucial reminder to consistently consider that chronic pelvic pain may originate from alternative sources in young women diagnosed with endometriosis.

Acknowledgements

We would like to thank all surgeons, the co-workers, the editor, and the reviewers for their assistance and guidance in this research.

Author contributions

All authors contributed to the study conception and design. M.R. performed the surgical procedure and designed the study. O.B., A.L., and K.B. collected the data. O.B. and H.S. wrote the first draft of the manuscript. Z.F. and S.D. examined the surgical specimen and ensured the final anatomopathological interpretation. Y.B. participated in the study design and critically reviewed the manuscript. M.M.A., F.S., H.A., and M.R. critically reviewed the manuscript. All authors read and approved the final version of the manuscript.

Data availability statement

All data is contained within the manuscript file.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethical approval

This case of report is only involving objective retrospective description therefore not applicable to ethics approval.

Informed consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

Trial registration

Not applicable.

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