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Giant Retroperitoneal Ganglioneuroma Successfully Resected with Preservation of the Kidney and Major Vessels: A Case Report

Mohamad Alharbi
Hama university
Ahmad Alabdlilrazzak
Hama university
Abd Albadee Abd Alkareem
Hama university
Mohammad Noor Katranjy
m8409842@gmail.com

Hama university
Samer Muhsen
Damascus University

Case Report

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Abstract

Background

Ganglioneuroma is a rare, benign tumor originating from the neural crest, with the retroperitoneum being the most common site of occurrence. It Is characterized by the absence of specific symptoms, as vague clinical manifestations predominate, usually due to the compression of adjacent organs. While radiological modalities such as CT and MRI are useful diagnostic tools, they are often insufficient due to the non-specific radiological features of the tumor, necessitating surgical intervention followed by histopathological study to confirm the diagnosis.

Case Presentation

We report the case of a young man who presented to the hospital with vague abdominal pain, a sensation of heaviness, and progressive abdominal enlargement. Clinical and laboratory examinations were within normal limits. Following ultrasound and CT scans, a massive retroperitoneal mass measuring 29.5 × 28 × 6 cm was detected. The patient underwent an exploratory laparotomy, which revealed a tumor originating from the lower thoracic vertebrae and extending into the pelvis, causing significant displacement of the left kidney and the major abdominal vessels. The tumor was successfully resected, and the patient was discharged three days postoperatively in excellent clinical condition.

Discussion and Conclusion

Although ganglioneuromas in the literature are typically smaller than 10 cm, this case is exceptional due to the tumor's massive size (29.5 cm). This report contributes to the medical literature by documenting an exceptionally rare presentation of a giant retroperitoneal ganglioneuroma that significantly exceeds typical dimensions, posing a substantial diagnostic and therapeutic challenge. It underscores the necessity of histopathology for a definitive diagnosis and confirms that surgical excision remains the definitive treatment with an excellent prognosis

Introduction

Ganglioneuroma is a rare neoplasm derived from neural crest cells; while It commonly localizes in the retroperitoneum, its origin from the adrenal medulla is significantly less frequent(1). These tumors are typically asymptomatic and frequently detected incidentally. Nevertheless, clinical manifestations may arise when the tumor reaches a significant size, causing compression of adjacent organs; such symptoms are generally non-specific, including abdominal pain, nausea, and vomiting. Although the tumor may secrete hormones like catecholamines or androgens in exceptional cases, such secretory activity is considered an extremely rare phenomenon.(2)

In most instances, Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) are Insufficient for a definitive diagnosis. Radiographic features, such as calcifications and a homogeneous appearance, are not pathognomonic, as they may mimic more aggressive malignancies—particularly when concurrent

with lesions in adjacent organs like the kidney and adrenal gland. Consequently, surgical intervention remains essential in most cases to establish a definitive histopathological diagnosis(3). The combination of vague non-specific symptoms, rarity, diagnostic imaging challenges, and clinical overlap with aggressive malignancies renders this tumor a diagnostic and therapeutic challenge that necessitates ongoing follow-up and further clinical study.

Case presentation

A 20-year-old Syrian male was admitted to Damascus University Hospital with progressive abdominal enlargement and vague abdominal pain for one year and abdominal heaviness sense.

Physical examination revealed a marked abdominal distension, no tenderness, no shifting dullness, other clinical findings were normal. Laboratory investigations were normal.

Abdominal Ultrasonography revealed a huge solid mass, computed tomography (CT) showing a well – circumscribed huge retroperitoneal tumor measures 29.5 x 28 x 6 cm (29.5 cm in the greatest dimension) .The tumor extends from thoracic vertebrae to the pelvic and displeased the left kidney and large abdominal arteries and veins (abdominal aorta, inferior vena cava). (Fig. 1)

The primary diagnosis suggested a differential between sarcoma and leiomyoma. The patient underwent an exploratory laparotomy via a midline incision. Intraoperatively, a mass arising from the inferior thoracic vertebrae and extending into the abdomen was identified. A Cattell–Braasch maneuver was performed to achieve adequate exposure, followed by a meticulous surgical procedure that lasted five hours to perform a complete radical resection. The tumor was accurately dissected from adjacent organs, including the left kidney and large abdominal vessels, necessitating IVC clamping and 20 minutes of renal clamping. To ensure vascular safety, intravenous Heparin was administered during the surgery, and any vascular repair was completed using 5/0 Prolene sutures. The removed retroperitoneal tumor measured 29.5 x 28 x 6 cm and was surrounded by a thin capsule (Fig. 2). The estimated blood loss was approximately 500 cc; consequently, one unit of blood was transfused intraoperatively and another unit was administered in the ward. Postoperatively, the patient remained in good clinical condition with no reported complications,Figure(1)

Histopathological examinations revealed benign spindle cell proliferation with mature ganglion cells, consistent of ganglioneuroma .The tumor is well-demarcated and surrounded with thin capsule. Surgical margins were free of tumor. Sections show a rubbery fatty cut-surface with necrotic areas. the final diagnosis is ganglioneuroma .

The patient was discharged after 3 days of surgical implementation in very good condition

Discussion

Ganglioneuromas represent the benign, fully differentiated end of the neuroblastic tumor spectrum; however, their indolent biology may be misleading from a surgical perspective. Large retroperitoneal lesions, as demonstrated in this case, can establish complex relationships with major vascular structures, resulting in significant technical challenges despite their non-invasive histology [1, 2].

Tumor size, rather than biological behavior, remains the primary determinant of surgical complexity. In this patient, the large mass caused marked displacement and apparent encasement of the inferior vena cava (IVC) and renal vein. Although preoperative imaging suggested a benign process, intraoperative findings revealed dense fibrotic adherence, highlighting the limitation of imaging In distinguishing true vascular invasion from firm adherence [1, 3]. Consequently, surgeons must anticipate difficult vascular dissection even In radiologically favorable cases.

An open approach via a midline laparotomy was adopted to ensure adequate exposure. A Cattell–Braasch maneuver enabled medial mobilization of the right colon and duodenum, facilitating direct access to the IVC and renal vessels. Given the tumor's proximity to major vasculature, early vascular control was established. Intraoperatively, clamping of the IVC and temporary occlusion of the renal vein for approximately 20 minutes were required to safely manage vascular injury. This period of venous clamping was well tolerated without postoperative renal impairment, consistent with previous reports supporting renal preservation under controlled conditions [2, 4].

Vascular injuries were managed with primary venorrhaphy using 5 – 0 polypropylene (Prolene) sutures. Preservation of vessel wall integrity allowed direct repair, avoiding complex reconstruction and minimizing operative morbidity. This approach aligns with contemporary vascular surgery principles favoring primary repair when feasible [5]. Estimated blood loss was approximately 500 mL, and hemodynamic stability was maintained with transfusion support.

While minimally Invasive approaches have been described for selected cases, their applicability remains limited in large tumors with major vascular involvement [7, 8]. In contrast, open surgery provides superior exposure and facilitates safe management of intraoperative vascular complications, as supported by systematic reviews of giant ganglioneuroma resections [6].

Finally, the management of such tumors underscores the importance of multidisciplinary preparedness. Retroperitoneal ganglioneuromas frequently surround or encase critical vascular structures, necessitating advanced surgical expertise and careful preoperative planning [1, 2].

In conclusion, radiological benignity does not equate to surgical simplicity. Large retroperitoneal ganglioneuromas should be approached with anticipation of vascular adherence and preparedness for vascular control and repair. With meticulous planning and intraoperative adaptability, complete resection with preservation of vital structures is achievable even in complex scenarios.

Declarations

Ethics approval and consent to participate:

Ethical approval was obtained from the institutional review board of Hama University.

Consent for publication:

Written informed consent was obtained from the patient for 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.

Competing interests:

The authors declare that they have no competing interests.

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Author Contribution

MA collected clinical data. MA, AA, and AK were responsible for writing and formatting the manuscript. MK handled the correspondence and submission process. SM performed the surgical procedure. All authors read and approved the final manuscript.

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Availability of data and materials:

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

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Figures

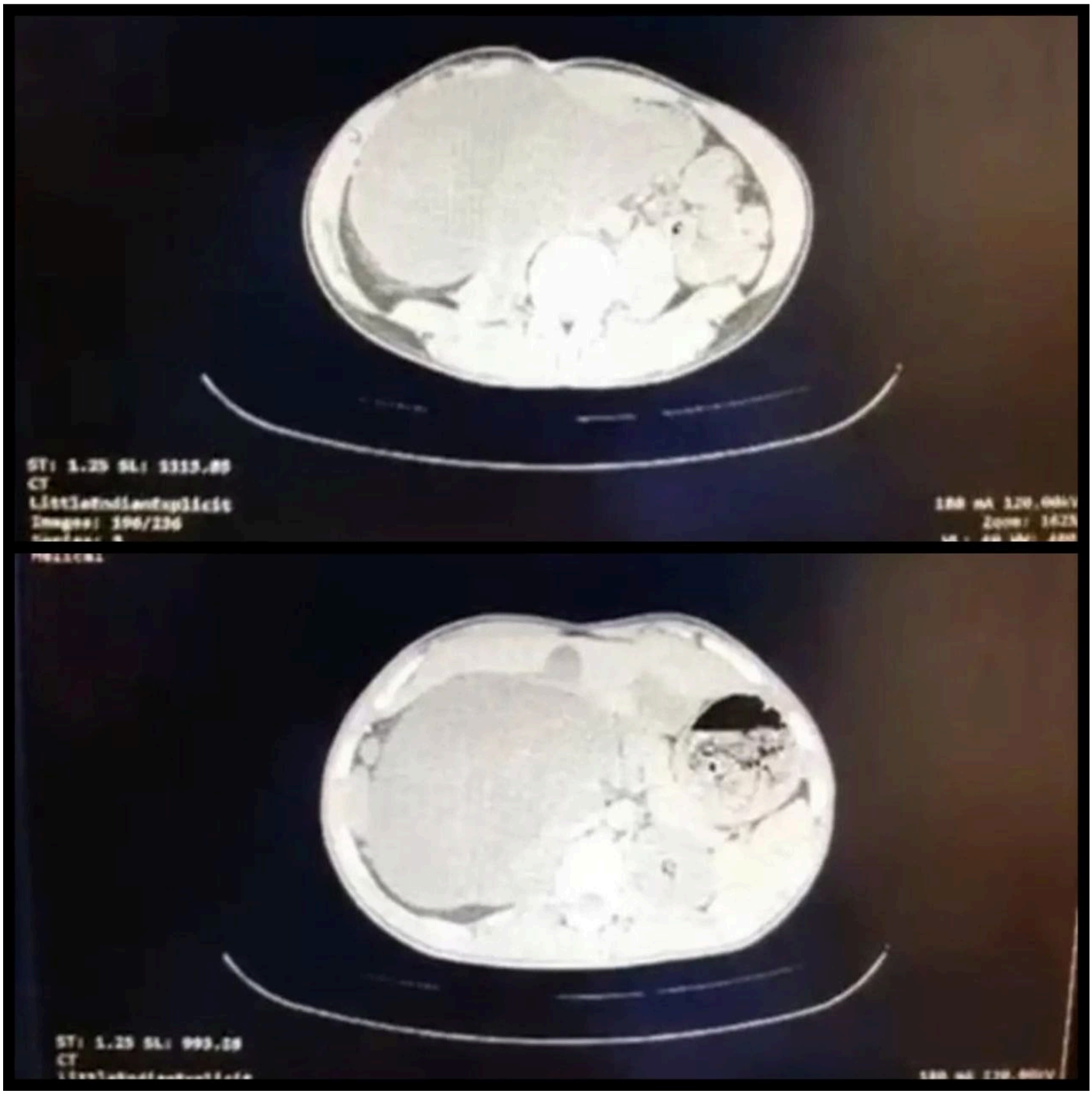
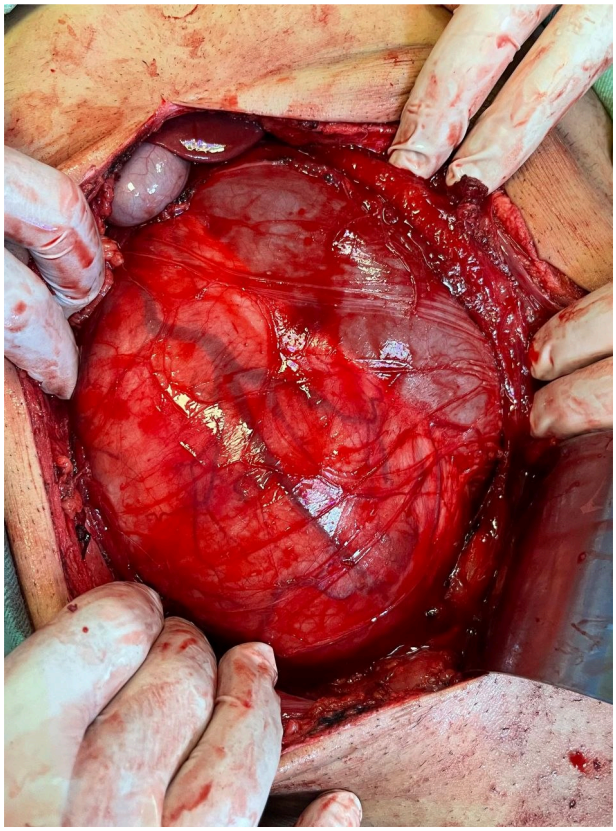
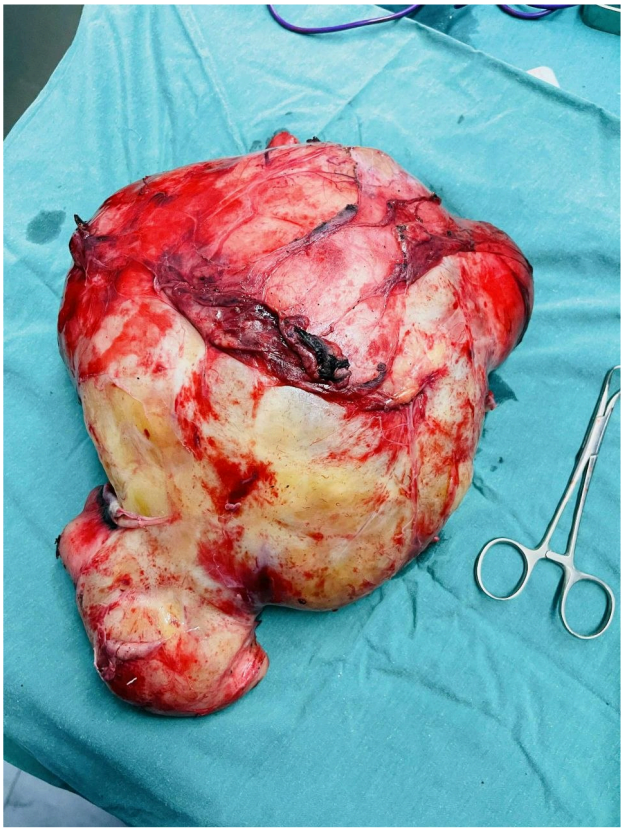


Figure 1

Axial CT images showing a 29.5 cm retroperitoneal mass



2a



2b

Figure 2

(a) Intraoperative view of the mass. (b) The resected 29.5 cm tumor specimen

a. Intraoperative view of the giant mass displacing major abdominal vessels.

b. The resected 29.5 \text{ cm} tumor specimen after successful total excision

Supplementary Files

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- Additionalfile1.mp4.mp4
- CAREChecklist.docx