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# Multidisciplinary management of chylous ascites following resection of a retroperitoneal, functional Paraganglioma

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ABSTRACT

Extra-adrenal, functional paraganglioma, if not medically controlled, requires early surgical resection. This can often be challenging when found in a retroperitoneal location, where resections are prone to lymphatic disruption and chyle leaks. Chylous ascites carry a significant risk of protein and caloric malnutrition, dehydration, and even mortality given the nutrient-rich characteristic of lost lymphatic fluid. Dietary modification involving minimal fat intake is often frontline conservative treatment, however, prolonged, heavy-volume chylous ascites will require advanced medical and surgical intervention. This case illustrates the difficulties, complexities, and solutions with multidisciplinary management of lymphatic leakage following resection of a functional, retroperitoneal paraganglioma.

## 1. Introduction

Pheochromocytoma and paraganglioma are rare catecholamine-producing neoplasms that arise in the adrenal medulla or extra-adrenal sites respectively.[1,2] Paragangliomas can arise anywhere in the sympathetic chain, with half found in the para-aortic region, between the diaphragm and inferior renal poles; given the risk of multicentricity, an extended vertical midline incision is recommended to approach the tumor and examine the entire paraspinal axis from the diaphragm to the pelvis.[3] The extent of such a dissection carries significant risk of complications typically found in other retroperitoneal surgeries. Retroperitoneal lymph node dissection or resection of the inferior vena cava (IVC) may disrupt lymphatic channels and lead to nutrient rich chylous ascites.[4] The incidence ranges from 1 to 9% following retroperitoneal node dissection and 1–5% following radical nephrectomy, and although rare, carries significant morbidity and mortality related to malnourishment and dehydration.[4,5] Dietary modification and placement of surgical drains are routine modalities of conservative management, however, parenteral nutritional support may be required for serious lymphatic leaks.[6,7] More invasive management, in the setting of prolonged or elevated chylous output, may require assistance from interventional radiology for lymphangiography with or without embolization, or even further surgical intervention.[5] We present a case of prolonged chylous ascites following retroperitoneal resection of a malignant, functional paraganglioma, requiring extensive nutritional support and endolymphatic intervention.

## 2. Case description

A 54-year-old woman presented to the urologic oncology clinic after an incidental finding of two retroperitoneal masses, measuring 3.9cm and 2.0cm, on a computed tomography (CT) scan of the abdomen for generalized abdominal pain. This was confirmed with positron emission tomography (PET) scan, demonstrating an FDG-avid retroperitoneal conglomerate with no additional sites of pathologic activity (Fig. 1). CT-guided biopsies, though complicated by hypertensive urgency, revealed adrenal medullary tissue consistent with pheochromocytoma. The patient consented to excision of the extra-adrenal, functional paraganglioma and was seen by cardiology, endocrinology, and medical oncology for optimization before surgery. Achieving adequate alpha- and beta-adrenergic blockade was prolonged and challenging. Admission to the intensive care unit (ICU) and initiation of intravenous

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nicardipine and esmolol finally yielded appropriate blood pressure control. Presurgical abdominal imaging demonstrated a conglomerate of three retroperitoneal nodal masses (Fig. 2): 4.1x3.3 × 6.9 cm infrarenal aortocaval mass with an inferior nodule at the aortic bifurcation (2.5x1.8 × 2.4 cm) and an additional aortocaval nodule (1.3 × 0.9 cm).

The patient successfully underwent induction of anesthesia and subsequent resection of the extra-adrenal, functional paraganglioma. Intraoperatively, the liver had a nodular appearance, however there were no ascites. The nodal conglomerate measured up to 10.5 cm with numerous parasitic venous tributaries, and was fully resected. The inferior vena cava was partially resected with the endovascular stapler at the site of engagement with the mass. Given the extent of the retroperitoneal dissection, there was concern for lymphatic disruption. Thus, a 19-French surgical drain was left in the resection bed, and they were transferred to the ICU for further postoperative management.

## 3. Complication event & management

On postoperative day (POD) 1, they were hemodynamically stable without vasopressors and transferred to the floor, where bowel function returned on POD2. The patient's diet was slowly advanced to a goal diet of zero fat by POD2. From POD2 to POD5, daily drain output was >1L and frankly chylous. Drain studies were performed, which were notable for elevated fluid triglyceride (801 mg/dL, [normal: <110 mg/dL]), and nucleated cell count (4893 cells/mL, [normal <500 cells/mL]), overall concerning for chylous ascites secondary to a lymphatic leak. The patient was maintained on a zero-fat diet and had strict fluids and diet monitoring. By POD7, drain output remained persistently high, and furosemide was administered for diuresis. Although drain fluid became less opaque, output remained high, and they were placed on <2L/day fluid restriction from POD11-12. During this time, they started supplementing their diet with low-to-zero-fat protein shakes, although the low protein-to-carbohydrate ratio was a concern given the risk of protein and caloric malnutrition in the setting of a lymphatic leak.

From POD13-16, daily drain outputs ranged from 2.7 to 3.7L, and the patient was further fluid-restricted to <1.5L/day. Given the persistent concern for lymphatic leak, a lymphangiogram via right iliac node access was performed by interventional radiology on POD17. There was accumulation of contrast among a potential nodal mass located at the L2-L3 level in the retroperitoneum (Fig. 3), however, upon retrospective review, leakage of the lipiodol (ethiodized oil) was observed. Although the chyle leak was minimally visualized and could be deemed diagnostic, there was no immediate therapeutic effect. On POD20, clamping trials every 6 hours were initiated to allow for fluid reabsorption and to mitigate nutritional losses. By POD23, there was growing concern for protein and calorie malnutrition, which prompted initiation of zero-fat peripheral parenteral nutrition (PPN). From POD24-30, full-day clamp trials were initiated until there was significant abdominal distension or pain endorsed by the patient, with daily volumes ranging from 0 to 900 mL. Lipids were added to the PPN in addition to starting home plant protein shakes containing only 1g of Fat and 20g of protein per serving, however, within a day their drain output began to reappear more chylous.

Given the extended duration of PPN with returning chylous ascites following attempted dietary advancement, the patient was started on total parenteral nutrition (TPN) on POD32. Repeat drain triglyceride was stable at 70 mg/dL, but without clinical improvement, the patient consented to a 2nd lymphangiogram on POD37. Scout radiograph demonstrated contrast pooling from the prior lymphangiogram across and along the inferior pelvic midline. Again, there was focal accumulation immediately to the right of the L3 vertebra, which was not safe for direct needle access. No contrast was seen superior to this point in the cisterna chyli, indicating a presumed lymphatic disruption at L3, however, without a sizable and straight-line lymphatic channel, embolization could not be performed safely (Fig. 4). Subsequently, drain output remained elevated with daily volumes 1–2L. Serial abdominal x-rays over the next 4 days demonstrated contrast extension up to the L2 vertebral body with layering along the lower pelvis from the previous lymphangiogram, consistent with the patient's chylous leak (Fig. 5).

By POD41, given the high output with persistent chylous ascites, TPN was maximally concentrated to just over 1L daily to maintain oncotic pressure. Drain outputs declined slightly to daily volumes of 500mL to 1.6L. Repeat fluid triglyceride was now 55 mg/dL, but in the context of elevated output, the concern for persistent lymphatic leak remained. The patient then consented to the third lymphangiogram on POD45 in addition to a transjugular liver biopsy with hepatic pressure assessment given the possibility of hepatic dysfunction in the setting of persistent ascites. The corrected sinusoidal pressure was 6 mmHg, not indicative of portal hypertension, while the liver biopsy demonstrated mild steatosis (10 %) with active steatohepatitis consistent with nonalcoholic fatty liver disease. Given the normal liver function tests throughout hospitalization, a hepatic etiology was very unlikely to be the source of the ascites. Lymphangiography was reattempted by injecting the target lymph node, however, this did not yield lymphatic duct access and the view was further obstructed by free-floating lipiodol in the peritoneum, leading the case to be aborted.

From POD46-56, drain output continued to gradually decrease from >1L to 50–475mL. On POD56, repeat CT of the abdomen/pelvis revealed near-complete resolution of free intraperitoneal fluid. On POD57, drain output was 50mL, and the decision was made to remove the drain. On POD59, a fourth and final lymphangiogram was reattempted, albeit unsuccessful secondary to obstructing view from free-floating lipiodol contrast. Given clinical and radiographic improvement in degree of ascites, they were discharged home on POD60 on TPN with scheduled follow-up with urology and endocrinology. Two months following discharge, the patient restarted and nicely tolerated an oral,

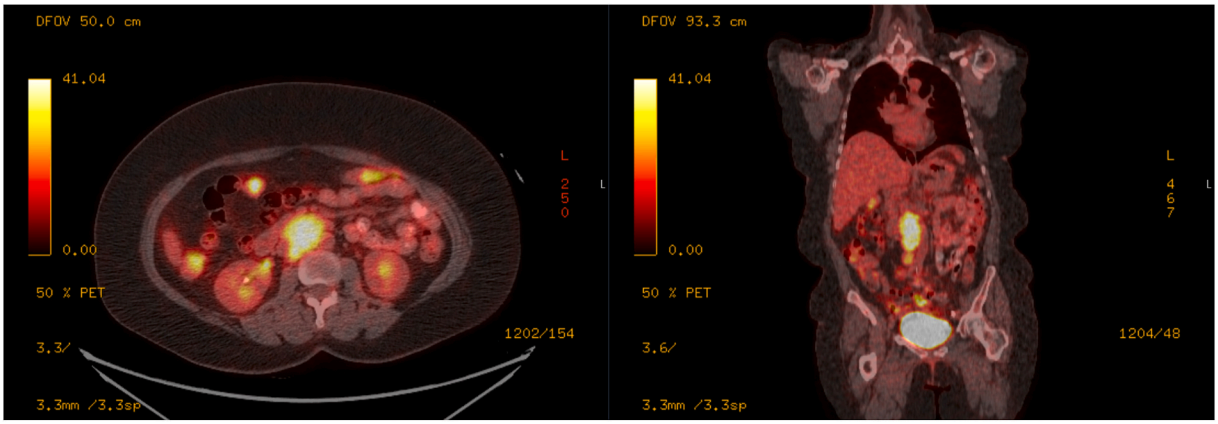


**Fig. 1.** Initial PET/CT scan demonstrating retroperitoneal nodal paraganglioma conglomerate at the level of the renal arteries.

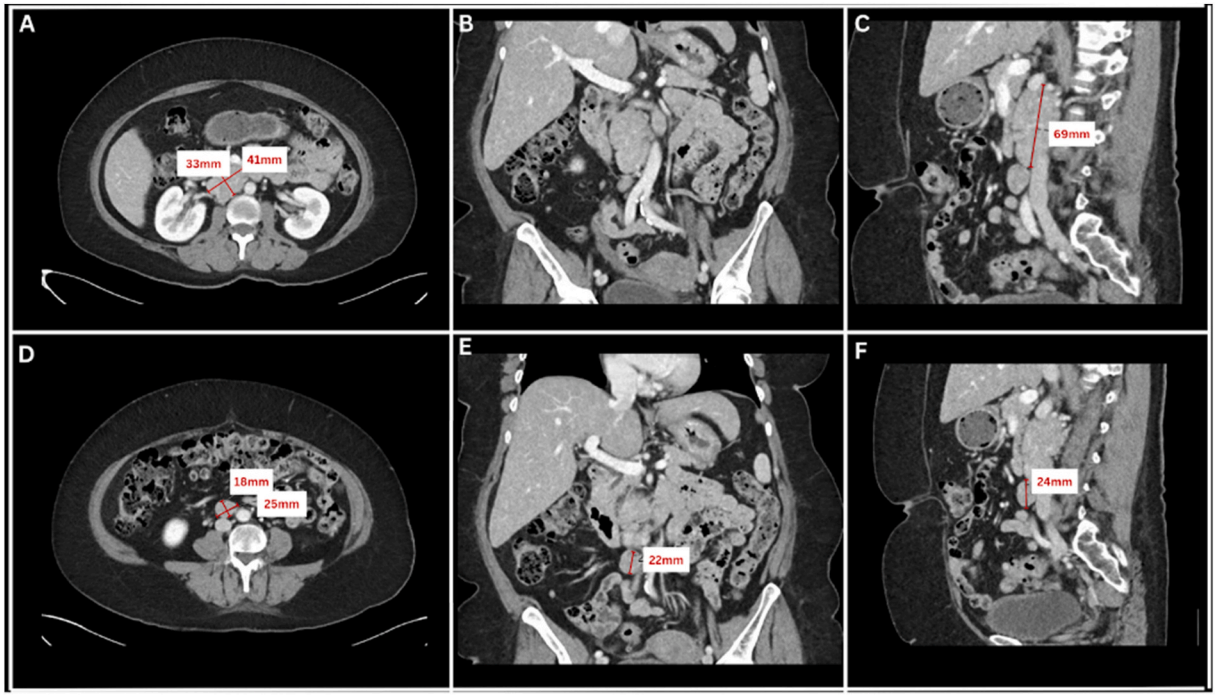


**Fig. 2.** Preoperative CT demonstration of paraganglioma conglomerate - (A) axial, (B) coronal, and (C) sagittal views of infrarenal aortocaval mass [4.1x3.3 × 6.9 cm], with (D) axial, (E) coronal, and (F) sagittal views of inferior nodule at the aortic bifurcation [2.5x1.8 × 2.4 cm].

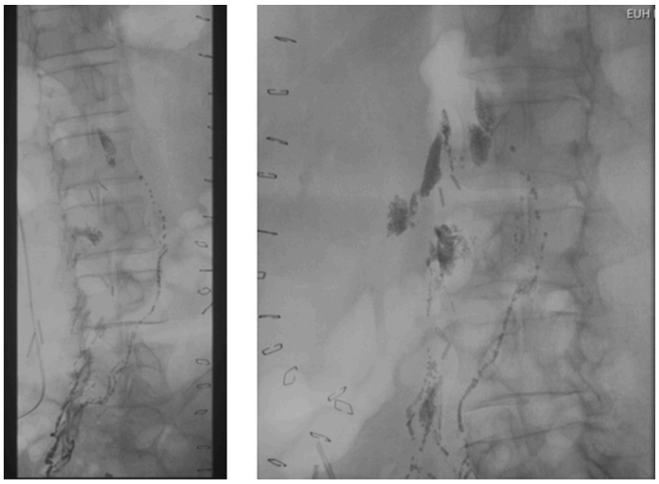

**Fig. 3.** POD17 Lymphangiogram #1 demonstrating accumulation of lipiodol contrast within retroperitoneal nodal mass at the L2-L3 level.

minimal-fat diet, and has since discontinued TPN. Energy has improved, they have returned to work, and have continued to regain weight as of their last outpatient visit, eight months following discharge. Repeat abdominal imaging demonstrated resolution of intra-abdominal fluid collections with unchanged distribution of contrast material related to prior lymphangiography. Final tumor pathology showed paraganglioma with 3 contiguous masses. Lymphovascular invasion was present, and the mitotic rate was >3 per 10 high-powered field. Most recent serum metanephrine and catecholamine markers were normal with no signs of residual or recurrent paraganglioma.

## 4. Discussion

Surgical resection is the mainstay treatment for functional paraganglioma, however, the present case represents the second published report of resection complicated by chylous ascites, with our patient managed via a combination of nutritional support and lymphangiography.[8,9] Lymphatic leaks can cause severe protein and caloric malnutrition, along with dehydration and other electrolyte deficiencies. Given the severity, early diagnosis is a priority, with surgical drains as important tools to monitor for occurrence or clinical resolution.[6,10] Once diagnosed, initial conservative management begins with a high-protein, medium-chain fatty acid (MCFA) exclusive diet, with emphasis on decreasing long-chain fatty acids given their packaging into chylomicrons.[5,7,11] This has been shown to decrease the rate of lymphatic flow by 200-fold, going from 200 mL/kg/hr to 1 mL/kg/hr.[5,11] If enteral nutrition fails to control the persistence or volume of drain output, parenteral nutrition is typically the next available option. PPN may be started via a peripheral intravenous line, however, TPN will require advanced intravenous access due to the hypertonicity and is reserved for patients in the setting of prolonged *nil per os* (NPO).[5,7] In addition, TPN has the benefit of being concentrated or volume reduced, while maintaining a greater percentage of amino acids, dextrose, and fats versus PPN, which is particularly useful when volume restriction is a concern. Dietary modification is typically sufficient for resolution of chylous ascites in over 60 % of postoperative patients, however, prolonged drainage for >21 days or excessive volume output >750 mL/day are indications more invasive treatment is needed.[5,12]

Historically, options for treatment of dietary refractory chylous ascites included surgical exploration with lymphatic ligation or peritoneovenous shunting for non-surgical candidates.[11,12] These procedures were highly morbid with significant complications.[5,11] With the introduction and improvement in intranodal lymphangiography, lymphatic imaging and intervention have exponentially advanced providing quicker, safer, and more effective methods of visualizing and treating lymphatic leaks.[13,14] Lipiodol, an ethiodized oil-based contrast agent, has widely been adopted in lymphangiography secondary to its diagnostic and therapeutic properties.[13,14] When lipiodol alone is injected, the contrast agent can extravasate at the site of lymphatic disruption, saponify the surrounding fat, and promote a granulomatous reaction that seals off the leak.[13,14] Among postoperative patients, lymphangiography alone with lipiodol can identify 55–67 % of patients

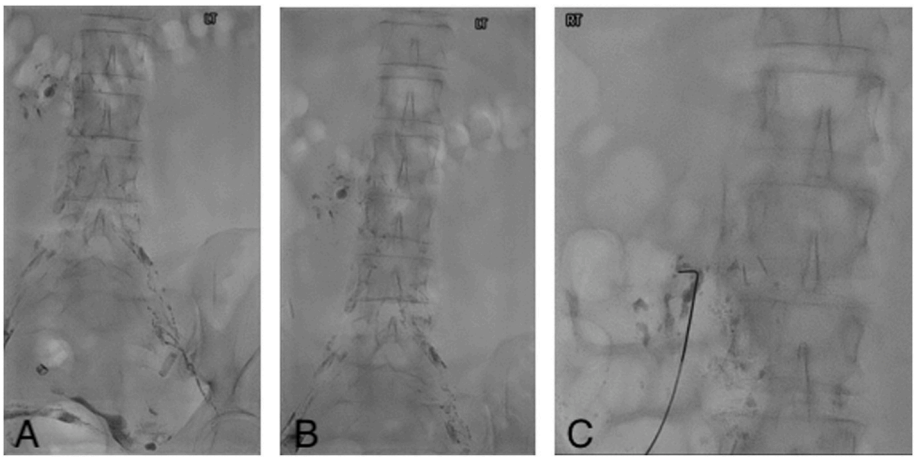


**Fig. 4.** POD37 Lymphangiogram #2 demonstrating prior contrast pooling across the inferior pelvic midline (A), with focal accumulation at the L3 level (B), failed attempt to access L3 collection directly (C).

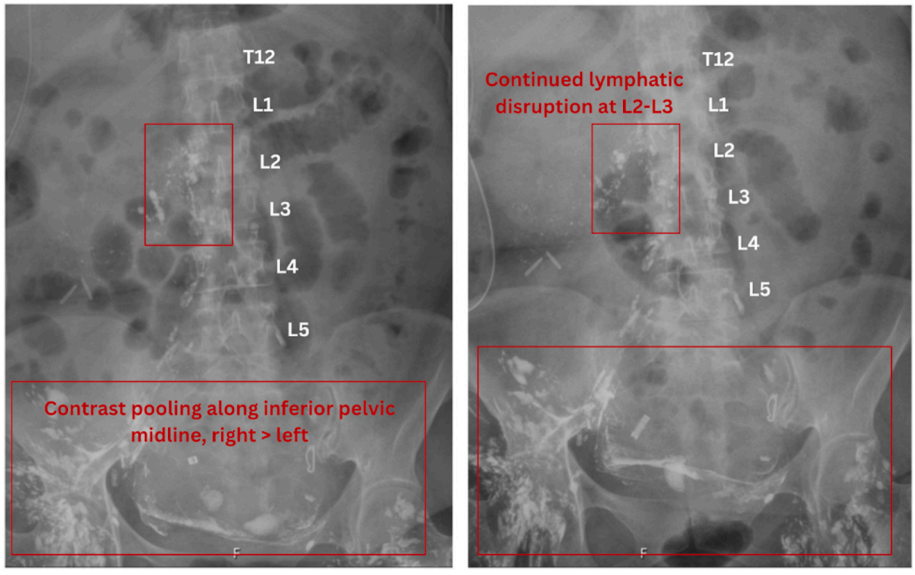


**Fig. 5.** Serial abdominal x-rays POD38-39 visualizing accumulation of contrast along the inferior pelvic midline consistent with a chylous leak.

with lymphatic leaks, with successful clinical improvement ranging from 35 to 70 %.[13,15–17] When lymphangiography alone is unsuccessful, adjunctive glue embolization is often employed, since lymphatics are fibrin and platelet-poor, allowing for glue to operate as a sealant.[14] The incorporation of glue embolization often improves success rate to 82–100 %, however, this includes patients requiring repeat intervention and was notably less successful among patients where no leak was ever identified (21 %).[13,15,16] In addition, glue embolization requires successful percutaneous access to lymphatics upstream from any identified leak, which can be complicated by the presence of lipiodol contrast from prior procedures, as seen in our case. Despite the challenges associated with lymphangiography and embolization, it carries fewer risks, and there is evidence to support repeated attempts of intranodal lymphangiography for the treatment of lymphatic leakage.[18]

## 5. Conclusions

Retroperitoneal surgeries can result in prolonged chylous ascites secondary to lymphatic disruption, requiring coordinated nutritional support and repeated interventions with lymphangiography. This case illustrates the complexity of modifying diet to account for nutritional losses from lymphatic leakage while considering how this may impact the prolongation or production of chylous ascites. Furthermore, this supports a role for repeat diagnostic and therapeutic lymphangiography to help manage refractory chylous ascites in a malnourished, post-operative patient, who may be unable to tolerate repeat surgical interventions.

## CRediT authorship contribution statement

**Edouard H. Nicaise:** Writing – original draft, Data curation, Conceptualization. **Ernest Morton:** Writing – review & editing. **Benjamin Croll:** Writing – review & editing. **Gregory Palmateer:** Writing – review & editing. **Gaurav Patel:** Writing – review & editing. **Bryan Swilley:** Writing – review & editing. **Viraj A. Master:** Writing – review & editing, Supervision, Conceptualization.

## Declarations

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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