



Laparoscopic Vena Cava Thrombectomy and Radical Nephrectomy in a Malignant Pheochromocytoma Case

Nefrectomia Radical com Trombectomia da Veia Cava Laparoscópica num Caso de Feocromocitoma Maligno

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Abstract

Pheochromocytomas with vena cava thrombus are extremely rare, with only a few cases reported in the literature. Radical nephrectomy with adrenalectomy and inferior vena cava (IVC) thrombectomy is the treatment of choice. However, it is a challenging procedure and its surgical approach is yet to be standardized. We present a case of a 49-year-old male incidentally diagnosed with a pheochromocytoma with aggressive local invasion and a level 1 vena cava thrombus. A laparoscopic right radical nephrectomy with right adrenalectomy, IVC thrombectomy and cavorrhaphy. A detailed revision of the technique is performed and compared with current strategies for pheochromocytoma optimal treatment. Renal and adrenal masses with vena cava thrombus are associated with high morbidity and mortality, particularly in the case of pheochromocytoma. The management is complex but minimally invasive surgery can be performed safely in the context of an experienced multidisciplinary team.

Keywords: Laparoscopy; Nephrectomy; Pheochromocytoma; Robotic Surgical Procedures; Thrombectomy; Vena Cava, Inferior

Resumo

Os feocromocitomas com trombo na veia cava são entidades extremamente raras, estando poucos casos descritos na literatura. Apesar da nefrectomia radical com adrenalectomia/suprarrenalectomia e trombectomia da veia cava inferior (VCI) corresponder ao tratamento de escolha, esta é uma técnica cirúrgica desafiante com uma abordagem ainda não padronizada. Apresenta-se um caso de um homem de 49 anos com diagnóstico incidental de um feocromocitoma localmente invasivo, com trombo na VCI nível 1. Foi proposta uma nefrectomia radical direita

laparoscópica com adrenalectomia/suprarrenalectomia, trombectomia da VCI e cavorrhafia. Neste artigo faz-se uma descrição detalhada da técnica cirúrgica e uma comparação com as estratégias atualmente utilizadas no tratamento do feocromocitoma. Tumores renais e suprarrenais/adrenais com trombo na veia cava estão associados a maior morbidade e mortalidade, sobretudo no caso do feocromocitoma. Apesar de complexa, a cirurgia minimamente invasiva é uma opção segura no contexto de uma equipa experiente e multidisciplinar.

Palavras-chave: Feocromocitoma; Laparoscopia; Nefrectomia; Procedimentos Cirúrgicos Robóticos; Trombectomia; Veia Cava Inferior

Introduction

Renal tumors with vena cava invasion and/or venous tumor thrombus are one of the most challenging conditions in urological oncology. It is not an infrequent finding in renal cell carcinoma, occurring in 4%-10% of cases, but it is extremely rare in adrenal pheochromocytomas. Radical nephrectomy with adrenalectomy and inferior vena cava (IVC) thrombectomy is the preferred treatment modality, improving both survival and quality of life.¹ Recently, laparoscopic and robotic assisted approaches have proven to be both oncologically safe and providing significant improvements in intra and post-operative surgical outcomes.^{2,3}

Our objective was to present a rare case of pheochromocytoma with aggressive local invasion and describe the surgical technique for tumor excision.

Case Report

Data was gathered by retrospective review of hospital clinical records.

A 49-year-old male patient presented with lipotimia, attributed to a diabetic ketoacidosis. Abdominal imaging showed an incidental 10 cm renal mass with extension to the ipsilateral adrenal gland, a level 1 vena cava thrombus and multiple enlarged para-cava, retro-cava and inter-aorto-cava lymph nodes (Fig. 1).

The patient underwent a right radical nephrectomy with concomitant right adrenalectomy, with en bloc excision of the right renal vein and a patch of IVC along with a level 1 thrombus. A retroperitoneal lymph node dissection was also performed. The

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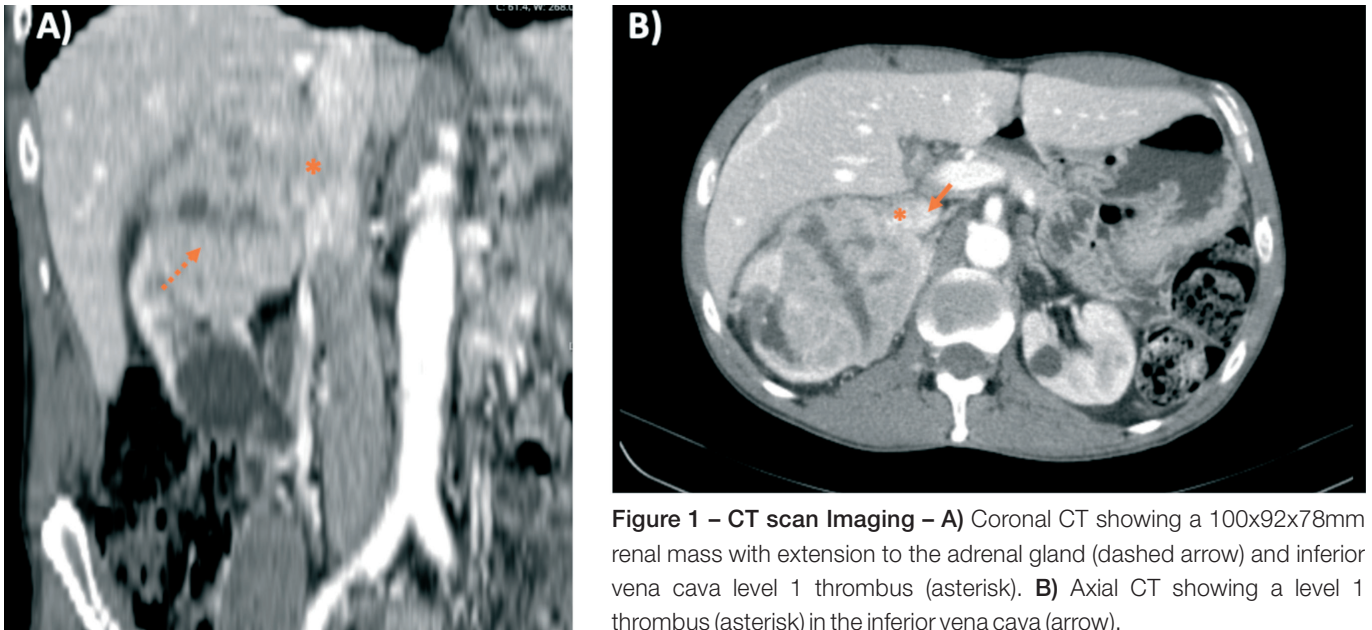


Figure 1 – CT scan Imaging – A) Coronal CT showing a 100x92x78mm renal mass with extension to the adrenal gland (dashed arrow) and inferior vena cava level 1 thrombus (asterisk). **B)** Axial CT showing a level 1 thrombus (asterisk) in the inferior vena cava (arrow).

patient was positioned in left lateral decubitus position and a 4 port transperitoneal access was performed, using a 12 mm paraumbilical trocar for a 3D telescope, one 12 mm and two 5 mm working ports. A transperitoneal approach was used and pneumoperitoneum was created with a 10 mmHg pressure. The right colon and the duodenum were mobilized along the Gerota's fascia plane. The vena cava was exposed and circumferentially isolated above and below the renal veins. Inter-aorta-cava lymph nodes were excised for safe vascular control. The right renal artery was identified and ligated at its origin. Control of both the vena cava, above and below the right renal vein, and the left renal vein was obtained with a double throw vessel loop tensioned with Weck Hem-o-lok non-absorbable polymer locking clips (Teleflex, Wayne, PA, USA). Clamping was performed sequentially, firstly at

the IVC below the renal veins, then the left renal vein and finally the IVC above the renal veins. Care was taken to avoid excessive renal vein and IVC manipulation. Cavotomy was performed and the right renal vein was liberated with the tumor thrombus and a patch of adjacent vena cava (Fig. 2).

The vena cava was closed with a running 4-0 PROLENE polypropylene (Ethicon, Somerville, NJ, USA) suture after appropriate flushing with heparinized saline to remove blood clots and gas bubbles (Fig. 3).

Sequential removal of the vessel loops revealed no leak in the suture. The remaining tumour mass, which was intimately adherent to the lateral and posterior vena cava wall, was carefully excised. Operative time was 260 minutes, blood loss was 400 cc (Fig. 4).

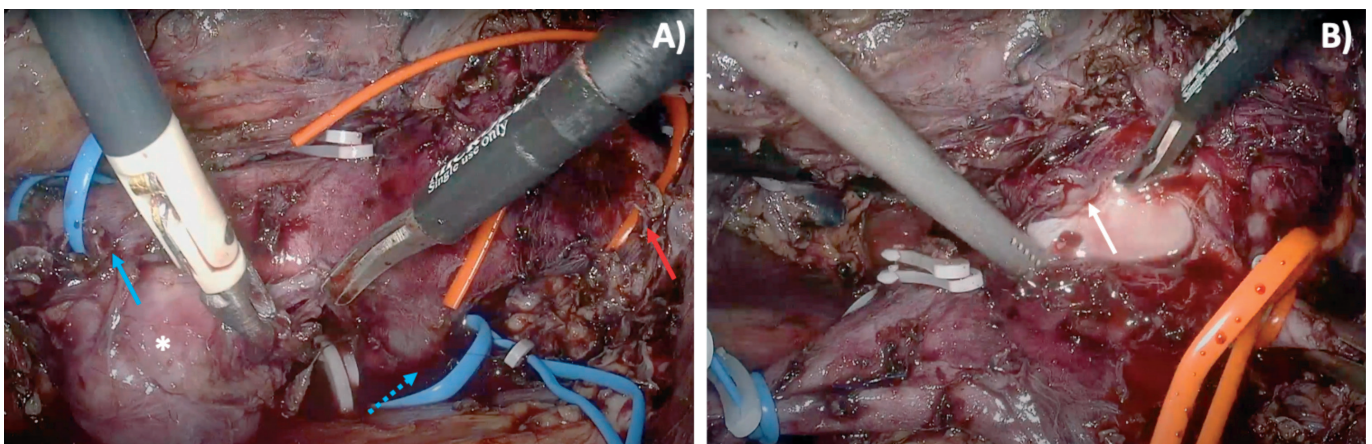


Figure 2 – Transperitoneal Laparoscopic approach – A) Vena cava and renal hilum control: suprarenal vena cava (red arrow), infrarenal vena cava (blue arrow) and left renal vein (dashed blue arrow) and inter-aorta-cava lymph node dissection (white www asterisk). **B)** Laparoscopic cavotomy and level 1 vena cava thrombus (white arrow).

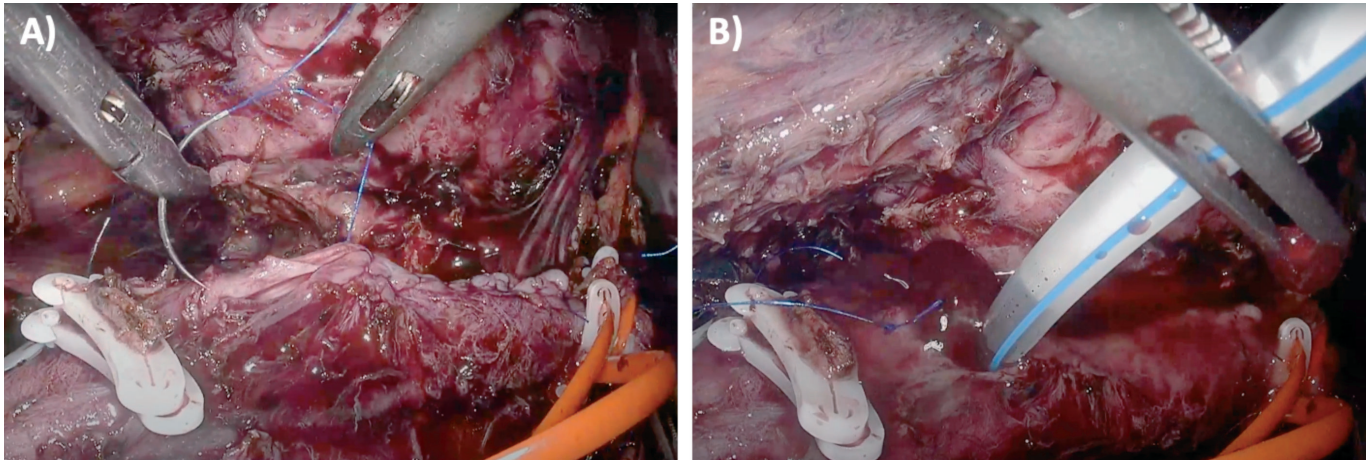


Figure 3 – Transperitoneal Laparoscopic approach – A) Cavourrhaphy with 4-0 polipropilene continuous suture. **B)** Flushing with heparinized saline.

The patient remained stable throughout the procedure, despite significant arterial tension variability. The postoperative period was uneventful, the drain was removed at postoperative day 2 with vestigial drainage and the patient was discharged at day 4. Pathology of the surgical specimen revealed a pT3 pN1 malignant pheochromocytoma. At 7 months follow-up the patient is asymptomatic, with normal tensional profile and no residual disease on postoperative computed tomography (CT) scan, requiring no adjuvant treatment at that time.

Discussion/Conclusion

Pheochromocytoma is a rare tumor with an annual incidence of 2 to 9 per million adults.⁴ Laparoscopic adrenalectomy has been the gold standard approach for the last few years.^{5,6} However, recent data have shown potential advantages of robotic-assisted surgery concerning obese patients and tumors larger than 5.5 cm.⁷

Nonetheless, open adrenalectomy is still a useful/valid approach, particularly for large potentially malignant or malignant tumors (>10 cm) and for those with organ infiltration.⁸ IVC thrombosis in these tumors may be caused by intravenous extension of the tumor, direct invasion through the vessel wall or by an hypercoagulable and inflammatory state associated with vessel injury.⁹ Nonetheless, intravenous extension is not a common feature in endocrine tumors.¹⁰ Therefore, the available evidence of pheochromocytoma excision with IVC thrombus is limited to a few case reports and a consensus on the optimal surgical approach as not yet been defined. Latest publications have shown a trend towards open surgery, however laparoscopic adrenalectomy, either via transperitoneal or retroperitoneal approach, have been successfully attempted.^{9,11-14}

Renal and adrenal masses with vena cava thrombus are associated with high morbidity and mortality, particularly in the

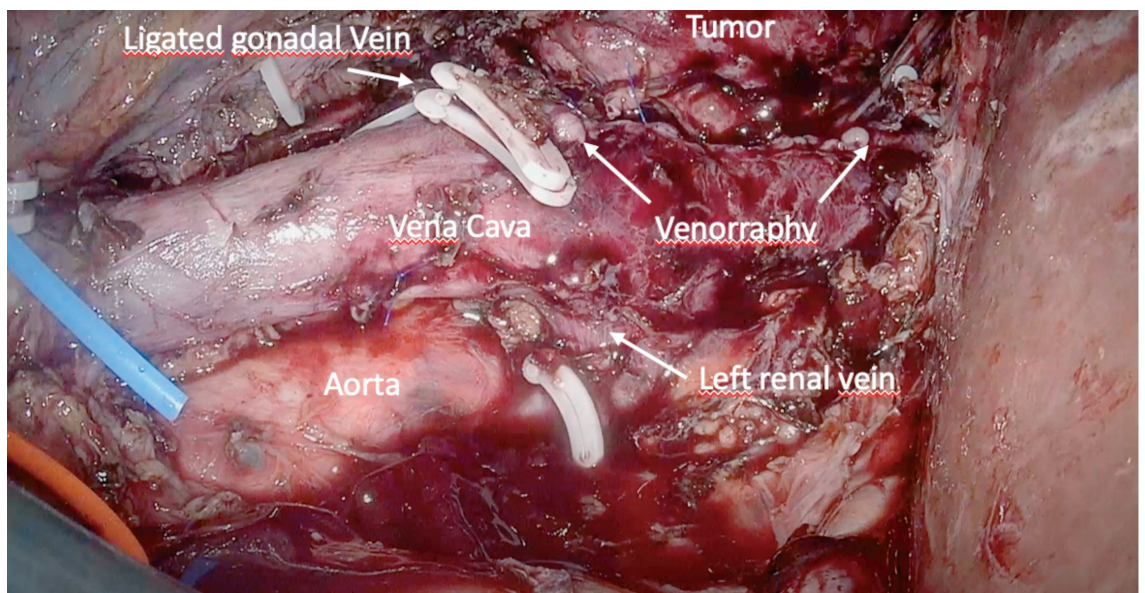


Figure 4 – Laparoscopic right nephrectomy and adrenalectomy with thrombectomy overview.



case of pheochromocytoma. Therefore, aggressive surgical resection is widely considered the treatment of choice for the management of these patients.¹ Despite current evidence showing no correlation between prognosis and thrombus level,¹⁵ thrombus site is an important marker for surgical technique and approach.¹⁶ Savage and Gill first reported a laparoscopic renal nephrectomy with renal vein tumor thrombus, using a gastrointestinal anastomosis stapler to isolate the distal renal and gonadal veins from the IVC.¹⁷ This procedure, however, is not adequate for level 2 thrombus or higher as it requires IVC clamping distally and proximally.

Early control of the renal artery previous to venal rein manipulation and kidney mobilization is a key step to minimize surgical complications.¹⁸ Hand-assisted laparoscopy was initially used for control of major vessels and kidney dissection.¹⁹

Postoperative embolic events, such as pulmonary thrombus embolism, is also a concern in IVC thrombectomy. IVC filters are not recommended on a standard basis as they can be associated with tumor infiltration and increased surgical complexity.²⁰⁻²² In our case instead, we followed the “IVC-first kidney-last” approach, developed by Chopra et al.²³ IVC dissection, with manipulation of surrounding tissues rather than the vein itself, exposure of the major vessels and early ligation before kidney mobilization reduces the chance of embolism and major hemorrhage.

Intraoperative ultrasonography has proved useful for assessing thrombus proximal extension, as in some cases preoperative imaging underestimates thrombus size and adherence to IVC wall.²⁴ In our case, laparoscopic ultrasound was not required due to good definition of tumor limits in preoperative CT. Furthermore, palpation of the IVC with the instruments showed it was fully collapsible, suggesting no tumor thrombus presence above the renal veins. However, it is recommended to have axial imaging close to surgery to confirm thrombus extension.

Pheochromocytoma excision carries an additional increased risk of intraoperative haemodynamic instability due to the release of catecholamines. A recent meta-analysis showed that laparoscopic excision of pheochromocytoma had a lower incidence of intraoperative haemodynamic instability events and overall complications when compared to open surgery.²⁵ The former findings might be explained by magnified laparoscopic field which allows for precise dissection and reduces unnecessary manipulation. However, these results must be weighed against the fact that tumor size might influence outcomes and that open surgery is usually performed on bigger tumors.

Improvements in imaging, surgical techniques and intraoperative monitoring enabled a decrease in perioperative mortality.²⁰ However, despite growing experience on renal tumors with venous thrombus and more advanced technology, such as laparoscopic and robot-assisted surgery, radical nephrectomy with IVC thrombectomy still remains a challenging procedure. There-

fore, adequate patient selection, technique standardization and multidisciplinary teams are of the foremost importance for minimally invasive surgery to be performed safely.

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