Laparoscopic excision of para-aortic ectopic pheochromocytoma

Jonathan M. Ng, MD,[1] Saleh Binsaleh, MD,[2] Britton Tisdale, MD,[2] Edward D. Matsumoto, MD,[1,2] Anil Kapoor, MD[1,2]

[1]Michael G. Degroote School of Medicine, McMaster University, Hamilton, Ontario
[2]Division of Urology, Department of Surgery, McMaster University, Hamilton, Ontario, Canada

NG JM, BINSALEH S, TISDALE B, MATSUMOTO ED, KAPOOR A. Laparoscopic excision of para-aortic ectopic pheochromocytoma. The Canadian Journal of Urology. 2006;13(5):3271-3274.

Ectopic pheochromocytomas (paragangliomas) are rare tumors that are traditionally treated with open surgery. We present our technique of such a tumor treated laparoscopically and review the safety, feasibility and advantages of this approach.

Key Words: ectopic pheochromocytoma, paraganglioma, laparoscopy, extra-adrenal pheochromocytoma

Background

Ectopic pheochromocytomas, also known as paragangliomas, are rare, catecholamine-secreting tumors arising from paraganglia cells of the autonomic nervous system.[1] Traditionally, open resection has been the recommended treatment approach.[2] With advances in minimally invasive surgery, paragangliomas may now be treated laparoscopically. However, relatively few cases have been reported and the evidence in support of the benefits and safety of the procedure is growing. We report our technique of laparoscopic excision of a para-aortic paraganglioma in a 34-year-old man who showed a 2.2 cm retroperitoneal soft tissue mass on abdominal and pelvis contrast CT.

Accepted for publication June 2006

Address correspondence to Dr. Anil Kapoor, Section of Urology, Department of Surgery, McMaster University, St. Joseph's Hospital, 50 Charlton Avenue East, Hamilton, Ontario, L8N 4A6 Canada

Surgical technique and case report

A 34-year-old caucasian man presented with hypertension and occasional palpitations, diaphoresis and headaches. He was initially treated with hydrochlorothiazide (Dyazide) and referred to nephrology. During workup of his hypertension, 24-hour urinary collection revealed elevated urinary norepinephrine (3699 nmol/d; normal, < 600 nmol/d) and urinary metanephrine (15.6 μmol/d; normal, < 5.5 μmol/d). Twenty-four hour ambulatory blood pressure monitoring revealed systolic blood pressure spikes of up to 170 mmHg. He was immediately started on alpha-1 adrenergic blockade with Prazosin and his hydrochlorothiazide was discontinued. Contrast CT scan of the abdomen and pelvis

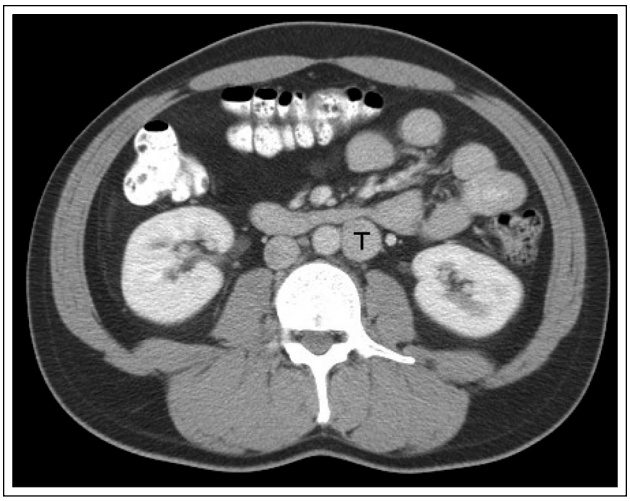

Figure 1. Abdominal contrast CT demonstrates a 2.2 cm mildly enhancing mass (T) in the retroperitoneum.

demonstrated a 2.2 cm mildly enhancing mass in the retroperitoneum with simple cysts in the left kidney, Figure 1. After experiencing dizziness, tachycardia and dry mucus membranes, the patient was placed on beta-blockers with Labetalol for symptom control. Iodine-131 metaiodobenzylguanidine (^{131}I-MIBG) scintigraphy showed no abnormal accumulation of tracer, Figure 2. T2 fat saturation MRI confirmed the

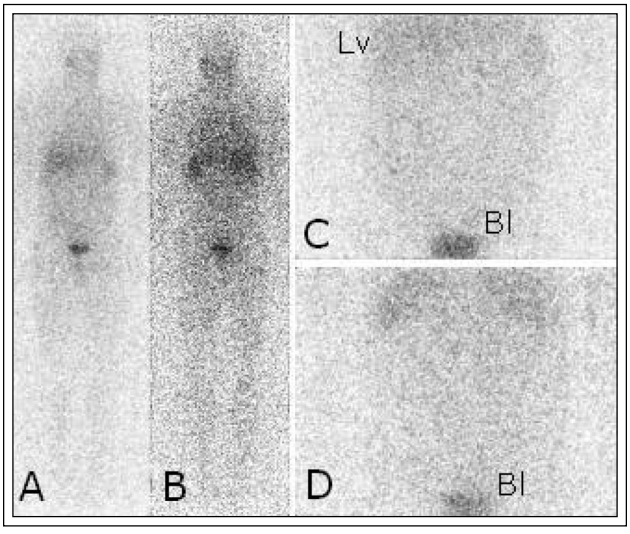

Figure 2. Iodine-131 metaiodobenzylguanidine (131I-MIBG) whole-body scintigraphy showing no abnormal accumulation of tracer 48 h after injection of the radiopharmaceutical: A) whole-body anterior view B) whole-body posterior view C) anterior abdominal view D) posterior abdominal view [Lv = liver, Bl = bladder]. 24 h studies were also negative (not shown).

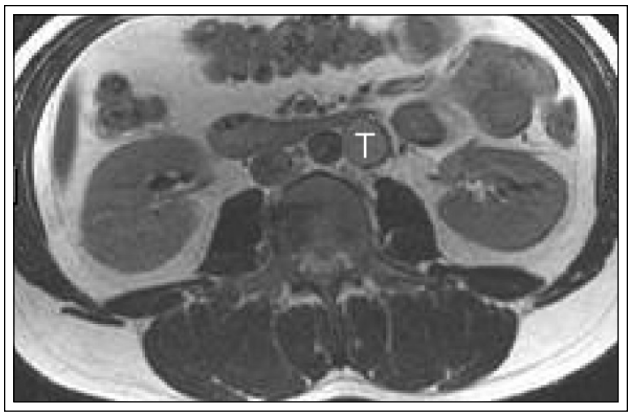

Figure 3. T2 Fat Sat MRI demonstrates a 2.6 cm solid hypervascular lesion (T) at the L3 level in the mid retroperitoneum just to the left of the aorta.

presence of a 2.6 cm solid hypervascular lesion at the L3 level in the mid retroperitoneum just to the left of the aorta, Figure 3. Interestingly, it did not have the classic "light bulb" sign of pheochromocytoma. He was placed on Phenoxybenzamine and the Prazosin was discontinued 3 weeks prior to surgery.

Under general anesthesia, pneumoperitoneum was established (15 mmHg) with insertion of the Verress needle at the umbilicus and three laparoscopic ports were inserted (2 mm x 10 mm, 1 x 5 mm). With regards to port position, Figure 4, the 10 mm camera port was

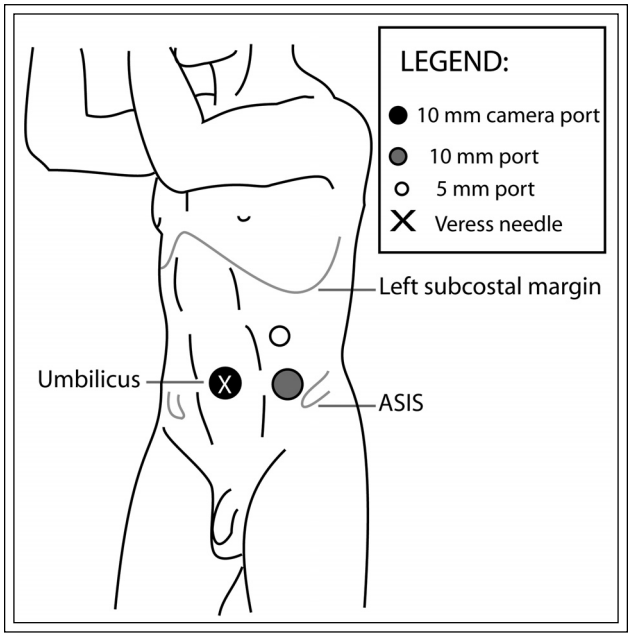

Figure 4. The port sites were clustered over the left upper quadrant to avoid adhesions and to allow the pneumoperitoneum to create adequate working space.

placed at the umbilicus, one 10 mm port placed halfway between the umbilicus and the anterior superior iliac spine (ASIS), and a last 5 mm port placed halfway between the umbilicus and left subcostal margin. After dissection of the white line of Toldt, the left ureter, gonadal vessel and psoas muscle were identified and preserved. The gonadal vein was followed superiorly to the left renal vein. The aorta was then identified and dissected laterally from this level down to the aortic bifurcation at the left common iliac artery. Anteriorly on the aorta, care was taken to preserve the Inferior Mesenteric Artery (IMA) and the Superior Mesenteric Artery (SMA). After this paraaortic dissection, the tumor was found just below the left renal hilum, Figure 5. The tumor was dissected carefully. Two arteries feeding the tumor from the aorta were clipped and divided. Next, three veins draining the tumor were also identified, clipped and divided. Following dissection, the specimen was placed into an EndoCatch® bag and withdrawn from a 10 mm port. The patient was sent to the recovery room without complication. Estimated blood loss for the procedure was about 10 cc and on postoperative day 1, hemoglobin levels were within normal range (149 g/L; normal, 130 g/L-180 g/L). The final pathology report concluded that the mass was a paraganglioma with no evidence of local invasion. Given the lack of local invasion into adjacent structures and the absence of detectable metastases, the tumor was concluded to be a benign paraganglioma. Postoperatively, the patient did very well and was discharged home on postoperative day 1.

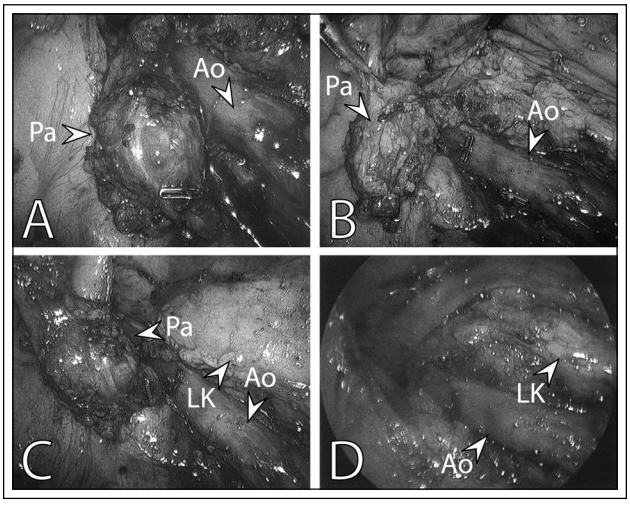

Figure 5. Intraoperative pictures. A) mass being dissected off of the aorta B) aortic dissection C) excision of paraganglioma mass D) completed dissection [Pa = paraganglioma, Ao = aorta, LK = left kidney].

Discussion

Ectopic pheochromocytomas are rare catecholamine-secreting tumors that occur outside of the adrenal glands.[1] Adrenal pheochromocytomas have an estimated incidence of around 1/100,000 and ectopic pheochromocytomas account for more than 10% of these cases in adults.[2-4] Extra-adrenal pheochromocytomas most commonly arise in the retroperitoneum, however, they may occur in the thorax, abdomen, pelvis, mediastinum and neck.[1,5] Clinical presentation of this condition consists of the cardinal signs and symptoms of catecholamine excess: headaches, diaphoresis, hypertension and palpitations.[1,6] Currently, the therapy of choice is surgical resection.[7]

Resection of ectopic pheochromocytomas must be performed with particular care. They are more likely multicentric, malignant and closely related to major vascular structures.[2,8] Compared to adrenal pheochromocytomas, ectopic pheochromocytomas present more often with multiple tumors. Approximately 28% of patients with ectopic pheochromocytoma present with multicentric tumors.[2,8] Historically, open surgical exploration and resection has been used to address these challenges.[2,9] However, advances in modern imaging techniques have overcome the need to perform full abdominal exploration.[10,11] Due to the accuracy of CT scanning, palpation of the retroperitoneum for additional tumors is no longer necessary. Also, preoperative ultrasonography and intraoperative laparoscopic ultrasonography may be used to search for additional tumors.[9,12] Another challenge to laparoscopic management of ectopic pheochromocytomas is their increased potential for malignancy. Rates of malignancy of up to 29%-40% has been reported in these tumors.[2,13-15] It was believed that an open approach would lead to better tumor control. However, we were unable to find any evidence in the literature to support this premise. Also, these tumors are often found in close relationship to major vascular structures.[2,8] Advances in laparoscopic technology, skill and technique have overcome these concerns. In the event of serious complications, the laparoscopic surgeon is able to convert the procedure to open surgery.

Laparoscopic surgery confers many benefits over open surgery. Previous studies comparing laparoscopic adrenal surgery have demonstrated faster recovery time, improved cosmesis, lower narcotic requirements and less pain.[11,16] Furthermore, recent reports support laparoscopic excision of ectopic pheochromocytomas as safe and effective.[7,9,17,18] According to one report, success with this approach can be attributed to careful preoperative medical

management, skillful intraoperative anesthesia and minimal adrenal gland manipulation.[7] We report our technique of successful laparoscopic excision of an ectopic para-ganglioma. Surgical technique caveats include careful attention to the SMA, IMA, and renal hilum, with care taken not to damage or divide these important structures often in close proximity to the ectopic mass. Feeding blood vessels to the ectopic mass often come straight off the aorta and must be carefully dissected without tearing to prevent aortic bleeding.

Given the available evidence, laparoscopic excision is likely a feasible, safe and advantageous approach to ectopic pheochromocytomas. Surgeons skilled in laparoscopic retroperitoneal surgery should consider this approach. Due to the rarity of this condition and lack of larger studies, collaboration between laparoscopic surgeons is recommended to investigate the long term outcomes of this approach. □

References

1. Yeo H, Roman S. Pheochromocytoma and functional paraganglioma. [review] [29 refs]. *Curr Opin Oncol* 2005;17:13-18.
2. Whitson BA, Tuttle TM. Laparoscopic resection of periaortic paragangliomas. *Am Surg* 2005;71:450-454.
3. Sheps SG, Jiang NS, Klee GG. Diagnostic evaluation of pheochromocytoma. [review] [43 refs]. *Endocrinology & Metabolism Clinics of North America*.1988;17:397-414.
4. Walz MK, Peitgen K, Neumann HP, Janssen OE, Philipp T, Mann K. Endoscopic treatment of solitary, bilateral, multiple, and recurrent pheochromocytomas and paragangliomas. *World J Surg* 2002;26:1005-1012.
5. Whalen RK, Althausen AF, Daniels GH. Extra-adrenal pheochromocytoma. [review] [156 refs]. *J Urol* 1992;147:1-10.
6. Townsend CM, Sabiston DC, MD Consult LLC. *Sabiston Textbook of Surgery: The Biological Basis of Modern Surgical Practice*. 17th / Courtney M. Townsend Jr. [et al. ed. Philadelphia: Elsevier Saunders; 2004.
7. Kercher KW, Novitsky YW, Park A, Matthews BD, Litwin DE, Heniford BT. Laparoscopic curative resection of pheochromocytomas. *Ann Surg* 2005;241:919-926.
8. Baghai M, Thompson GB, Young WF,Jr, Grant CS, Michels VV, van Heerden JA. Pheochromocytomas and paragangliomas in von hippel-lindau disease: A role for laparoscopic and cortical-sparing surgery. *Arch Surg* 2002;137:682-8;discussion 688-689.
9. Hwang J, Shoaf G, Uchio EM et al. Laparoscopic management of extra-adrenal pheochromocytoma. *J Urol* 2004;171:72-76.
10. Orchard T, Grant CS, van Heerden JA, Weaver A. Pheochromocytoma—continuing evolution of surgical therapy. *Surgery* 1993;114:1153-1158.
11. Walther MM, Keiser HR, Linehan WM. Pheochromocytoma: Evaluation, diagnosis, and treatment. [review] [47 refs]. *World J Urol* 1999;17:35-39.
12. Pautler SE, Choyke PL, Pavlovich CP, Daryanani K, Walther MM. Intraoperative ultrasound aids in dissection during laparoscopic partial adrenalectomy. *J Urol* 2002;168:1352-1355.
13. Jalil ND, Pattou FN, Combemale F et al. Effectiveness and limits of preoperative imaging studies for the localisation of pheochromocytomas and paragangliomas: A review of 282 cases. French association of surgery (AFC), and the French association of endocrine surgeons (AFCE). *Eur J Surg* 1998;164:23-28.
14. Melicow MM. One hundred cases of pheochromocytoma (107 tumors) at the columbia-presbyterian medical center, 1926-1976: A clinicopathological analysis. *Cancer* 1977;40:1987-2004.
15. Remine WH, Chong GC, Van Heerden JA, Sheps SG, Harrison EG Jr. Current management of pheochromocytoma. *Ann Surg* 1974;179:740-748.
16. Guazzoni G, Montorsi F, Bocciardi A et al. Transperitoneal laparoscopic versus open adrenalectomy for benign hyperfunctioning adrenal tumors: A comparative study. *J Urol* 1995;153:1597-1600.
17. Janetschek G, Finkenstedt G, Gasser R et al. Laparoscopic surgery for pheochromocytoma: Adrenalectomy, partial resection, excision of paragangliomas. *J Urol* 1998;160:330-334.
18. Ponsky LE, Gill IS. Laparoscopic excision of suspected extra-adrenal pheochromocytoma located in the mesenteric root. *J Endourol* 2002;16:303-305.